

Textbook of ORAL MEDICINE

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Textbook of ORAL MEDICINE

Second Edition

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Textbook of Oral Medicine

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*To
My Mother,
My Daughter Milini,
My Son Sanvil,
My Wife Savita
and
My Family*

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Foreword

“Enthusiasm is a driving force that overcomes all obstacles”

It is my proud privilege to write a foreword for second edition of this book by Dr Anil Ghom. In short time, this book has become most popular among undergraduates and postgraduates all over the country.

In the second edition, numbers of chapters have been presented in a better organized manner. This book carries updated information of the subject in this rapidly changing world of science. A new chapter “Controversial Diseases and Terminologies” is incorporated, also there are large number of new photographs, radiographs, MCQs and references. I am sure this new updated second edition will be more beneficial for the undergraduate and postgraduate students for reference and regular reading.



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Preface to the Second Edition

'We become strong only after we have acknowledged our weakness. Gather knowledge, insight, and experience and then make your own decision'

This is the second edition of the textbook and I am gratified by the acceptance and support that the book has received over the years from educators, students and practitioners. The purpose of second edition of this book is two-fold. First, to include what is new and recent knowledge and the second is correct shortcoming of the first edition. I have evaluated and utilized suggestions from all the critical reviews and recommendations from the faculty members.



Obviously, each successive edition of textbook finds the edition with more information. So in this edition I have attempted to solidify and include recent knowledge. This book also includes a new Chapter *Controversial Diseases and Terminologies*. Purpose of this chapter is that as knowledge is changing everyday with advanced diagnostic techniques, many old terminologies are discarded and new one are introduced.

My first book was criticized a lot for not including references and not having enough photographs in the book. So in this second edition I have included references and around 1000 photographs/illustrations for easy understanding of the diseases.

MCQs chapter is completely revised and all the new MCQs are added at the end of chapter. I have also included diagnosis of each lesion in diseases, so that students can understand key point in disease and they can easily remember it.

Again, as a human being, mistakes are bound to happen. I tried this second edition to best of my efforts, still there can be shortcoming and I request readers to make note of it and I will try to rectify it in the next edition.

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Preface to the First Edition

"You must not be discouraged if the world does not rush to you, demanding what you have; neither must you quietly sit down to let world wonder and then seek you; you must be aggressive, you must carry your truths to people and cause them to see them so clearly that they must accept them"

The student looked for a reference on which to build an educational foundation with regard to basic principle. A few years ago much of the information offered in this text was not available.

Since the principle and treatment modalities offered herein will continue to evolve, it behooves the student to be fully informed of the state-of-art to be able to critically evaluate the worthiness or applicability of any future development.

I have endeavored to ensure that a consistent style has emerged and is in harmony where appropriate with the diseases of oral region along with the differential diagnoses which are covered in detail.

The purpose of this book is to correlate the gross and microscopic pathological features with the radiographic appearance of oral diseases and systemic diseases manifested in the jaw.

In our increasingly litigious society, it is vital that the dentist understands the law as it relates to dentistry. The Chapter on Medicolegal Issue is Essential Reading, along with the Consumer Protection Act.

Diseases can be understood best when the interpreter understands not only the disease process but also the basic science associated with it. For this reason, I have included separate section for basic science.

Recently, as exam pattern is changing and MCQs are getting importance, MCQs are added in separate chapter.

I tried my best, to cover all the aspects of oral diseases in my book. If this goal is achieved, then this textbook may contribute, in a small way to better care of patients who suffer from these diseases.



Anil Govindrao Ghom

Acknowledgements

‘The man who really wants to do something finds a way, the other finds an excuse’

No work will be complete without help of your friends and well wishers and I am lucky to have them with me in this venture.

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Beauty is God’s gift but to utilize it in proper direction is in your hand. I am thankful to all those beautiful faces in book— Dr Gagandeep Chawala, Dr Smiriti Goswami, Dr Uma Rohra, Dr Swapnil Dewakirti, and Dr Pragya Jaiswal.

I would like to thank Dr Ranit Chhabra, Dr Priyanka Aggarwal, Dr Shaheen Hamdani, Dr Payal Tapadiya, and Dr Vivek Lath for their help in proofreading.

I offer my humble gratitude to my guide Dr RN Mody for his guidance during my postgraduation and after post-graduation.

Friends are always big supporters so I am grateful to Dr Pravin Sundarkar, Dr Neeraj Alladwar and Dr Ravindra Govindwar for heartily wishes. I am also thankful to my brothers and sisters especially elder brother Sadanand for their moral support in my life.

I am thankful to Shri Jitendar P Vij (Chairman and Managing Director) and Mr Tarun Duneja (Director-Publishing) of M/s Jaypee Brothers Medical Publishers (P) Ltd., for publishing this book. Commendable type setting, proofreading and improvement in illustrations have been done respectively by Ms Sunita Katla, Md Shakiluzzaman & Ms Geeta Srivastava and Mr Sumit Kumar.

Whenever I think who completely changes my life is my wife Savita, who is there in my thick and thin. Whenever I am down, she is there to uplift me with her prayer and support. She is a person with generous heart and I am thankful to God to give gift like her to me in my life.

Lastly, I offer my earnest prayers to the Almighty for endowing me the strength and confidence in accomplishing to the best of my abilities.

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Section **1**

Basics



Oral Diseases: An Introduction

The ultimate aim of entire dental education is to see how well it prepares the practitioners to serve patients. If one has to be a good practitioner it is essential to have a thorough understanding of the basic sciences related to dentistry.

Stomatology is the science of structure, function, and disease of the oral cavity. Study methods include examination of related histories, evaluation of clinical signs and symptoms and use of biochemical, microscopic and radiological procedures to establish a diagnosis and a plan for therapeutic management.

Diagnosis is the process of evaluating patient's health as well as the resulting opinions formulated by the clinician. *Oral diagnosis* is the art of using scientific knowledge to identify oral disease processes and to distinguish one disease from another.

History of oral medicine starts when *William Gies* of Columbia University in 1926 recommend that oral medicine topics should be covered in dental curriculum. In 1945, the American Academy of Oral Medicine was formed. Oral medicine definition accepted in 1993 by international association of oral medicine. It states that —

'Oral medicine is that area of special competence in dentistry concerned with diseases involving the oral and paraoral structures. It includes the principles of medicine that relate to the mouth as well as research in biological, pathological, and clinical spheres. Oral medicine also includes the diagnosis and medical management of diseases specific to the orofacial tissues and oral manifestations of systemic diseases. It further includes the management of behavioral disorders, the oral and dental treatment of medically compromised patients'.

It can also be defined as 'diagnosis and treatment of oral lesions as well as non-surgical management of temporomandibular joint disorders and facial pain and dental treatment for medically compromised patients in an outpatient sitting, or in an inpatient sitting under general

anesthesia, including specialty care in periodontics and endodontic'.

The goal and objective of oral medicine are discussed below. The goal is to provide education, research and service for health care professionals and the public.

- *Education*—it consists of predoctoral, postdoctoral and continuing education training for the health care professional.
- *Research*—it includes activities in the field of biology as it is related to oral disease.
- *Service*—service to society and health care professionals is the objective of oral medicine. Oral medicine will train the professional to provide current and future patient care.

As nowadays, epidemiology is changing, in the future, oral medicine person has to come across many oral diseases and he has to diagnose them. World Health Organization in 1989 study called 'trends in oral health care, a global perspective' told that in future greater role is required by the oral medicine professional.

In the field of oral medicine, you should have a basic understanding of various diseases and their impact on oral tissue, so that it is easy for a practitioner to recognize the presence of any major systemic diseases and then accordingly make the correct diagnosis and treatment plan so as to do thorough justice of what is happening to him.

The field of oral medicine consists chiefly of the diagnosis and medical management of the patients with complex medical disorders involving the oral mucosa and salivary glands as well as orofacial pain and temporomandibular joint disorders. Specialists trained in oral medicine also provide dental and oral health care for patients with medical diseases that affect dental treatment, including patients receiving treatment for cancer, diabetes, cardiovascular diseases, and infectious diseases (**Fig. 1-1**).

Oral medicine practice provides physical and medical evaluation, head and neck examination, laboratory

analysis, oral diagnosis and oral therapeutics for such conditions as: vesiculobullous, ulcerative mucosal diseases, painful and burning mucosa, infectious oral diseases, oral conditions arising from medical treatment, oral manifestations of systemic diseases and salivary gland dysfunction.

The specialist will perform a comprehensive and/or specialized examination, provide consultation, possibly perform and interpret laboratory tests and perform or prescribe treatments or make the appropriate referrals.

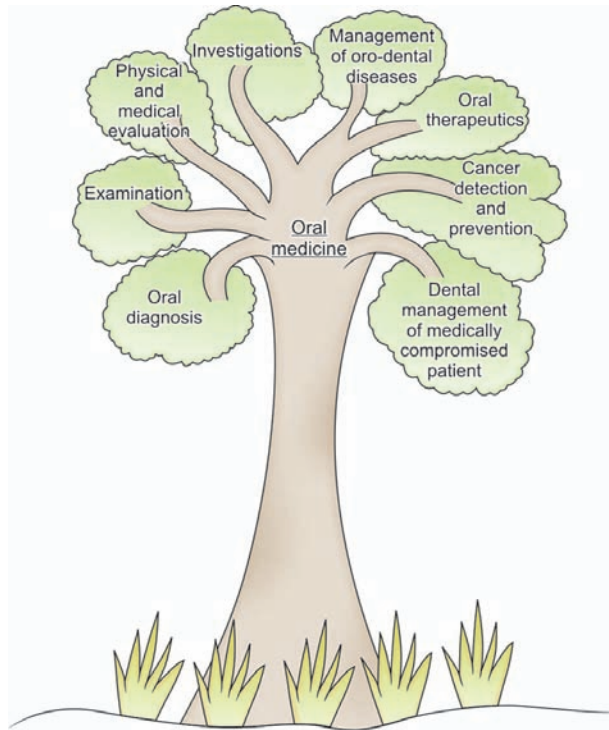


Fig. 1-1: Diagrammatic representation of different areas of oral medicine showing branches and goal of oral medicine.

Dental management of medically compromised patients is becoming a routine and increasingly important part of dental practice. Several factors contribute to this phenomenon. First, the population continues to age. Many older patients have multiple medical conditions. Second, as medical care becomes more effective and cost issues are emphasized, many patients are being treated on an ambulatory basis to avoid hospitalization. Consequently, these individuals are in the community and readily seek dental care. Third, the sophistication of medical treatment is prolonging life. And fourth, the level of and access to available dental care has improved, resulting in more patients (regardless of medical status) wanting dental treatment. Therefore, behavior disorders and diseases of the mouth as manifestations of systemic disease are seen at an increasing rate, and require prompt and adequate care by experienced specialists.

Philosophically and in practice, dentistry is similar to one of the various specialties of medicine and consequently,

it is imperative that the dentist understands the medical background of patients before beginning dental therapy, which might fail because of the patients compromised medical status or result in morbidity or death of the patients.

The dentist trained in oral medicine should be philosophically atuned to the patient and have knowledge of medically important diseases as well as of dental problems. The dentist should be well versed in the use of rational approaches in diagnosis, medical risk assessment and treatment.

The hospital is frequently the setting for the most complex cases in oral medicine. Hospitalized patients are most likely to have oral or dental complications of bone marrow transplantation, hematological malignancies, poorly controlled diabetes, major bleeding disorders, and advanced heart disease. The hospital that wishes to provide the highest level of care for its patients must have a dental department.

The hospital dental department should serve as a community referral center by providing the highest level of dental treatment for patients with severe systemic disease and management of the most medically complex patients is best performed in the hospital because of the availability of sophisticated, diagnostic and life-sustaining equipment and the proximity of expert consultants in all areas of health care.

Most difficult and unusual problems evaluated by the dentist are seen as consultations. To handle consultations properly, the dentist must be familiar with the proper method of requesting and answering consultations.

The role of imaging in oral medicine varies greatly with the type of problem being evaluated. Certain problems, such as pain in the orofacial region, frequently require imaging to determine the origin of the pain. For other conditions, however, such as soft-tissue lesions of the oral mucosa, imaging offers no new diagnostic information.

Thus, to conclude oral medicine expert is an important professional in dental and medical team of nations health care scheme to public. Oral medicine personal is also expert in studying, diagnosing and treating the mouth disease.

Suggested Reading

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2

Development and Eruption of Teeth

Introduction

Development of tooth is a result of complex process occurring between oral epithelium and underlying mesenchymal tissue. The primitive cavity is lined by stratified squamous epithelium, i.e. oral ectoderm, which contacts the endoderm of foregut to form the buccopharyngeal membrane. At 27th day of gestation, this membrane ruptures and primitive oral cavity establishes a connection with the foregut.

The scale of human tooth development:

- 42-48 days—dental lamina formation.
- 55-56 days—bud stage—deciduous incisor, canine and molar.
- 14th week—bell stage for deciduous bud for permanent.
- 18th week—dentin and functional ameloblast.
- 32nd week—dentin and functional ameloblasts of permanent 1st molar.

Stages of Tooth Development

Dental Lamina Formation

- *Proliferation of basal cells*—proliferation of certain areas of basal cells of the oral ectoderm occurs more rapidly than cells of adjacent area. This will result in formation of dental lamina. Dental lamina is a band of epithelium which has invaded underlying ectomesenchyme along each of the horseshoe shaped future dental arch (**Fig. 2-1**).
- *Time taken for dental lamina formation*—total activity of dental lamina formation extends at least over a period of 5 years. The remnants of dental lamina persist as epithelial pearls or islands within the jaw as well as in the gingiva.
- *Successional lamina*—the lingual extension of dental lamina is called as successional lamina.

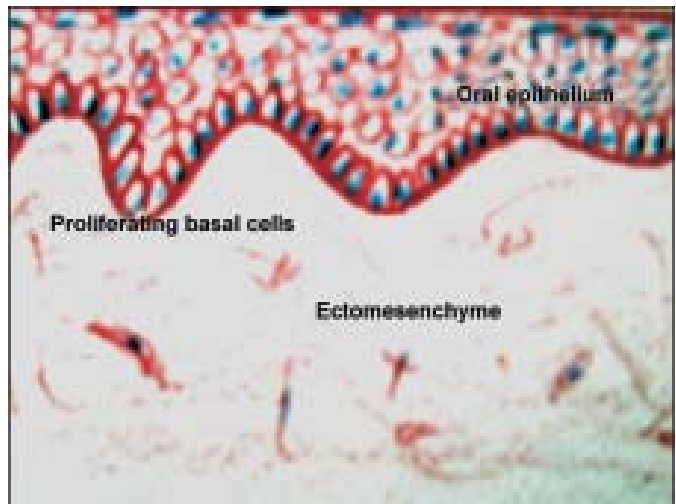


Fig. 2-1: Schematic diagram showing initiation of dental lamina by some proliferating basal cells.

Bud Stage (Initiation)

- *Primordia of enamel organ*—after the differentiation of dental lamina, round or ovoid swellings arise from basement membrane. This arises at 10 different points which corresponds to the future position of deciduous teeth. These are primordia of the enamel organs, the tooth bud.
- *Enamel organ*—the enamel organ consists of peripherally located low columnar cells and centrally located polygonal cells.
- *Dental papilla*—the area of ectomesenchymal condensation immediately adjacent to enamel organ is called as 'dental papilla'. Cells of dental papilla form future tooth pulp and dentin (**Fig. 2-2**).
- *Dental sac*—the condensed ectomesenchyme that surrounds the tooth bud and dental papilla is called as a 'dental sac'. Cells of dental sac form cementum and periodontal ligament.

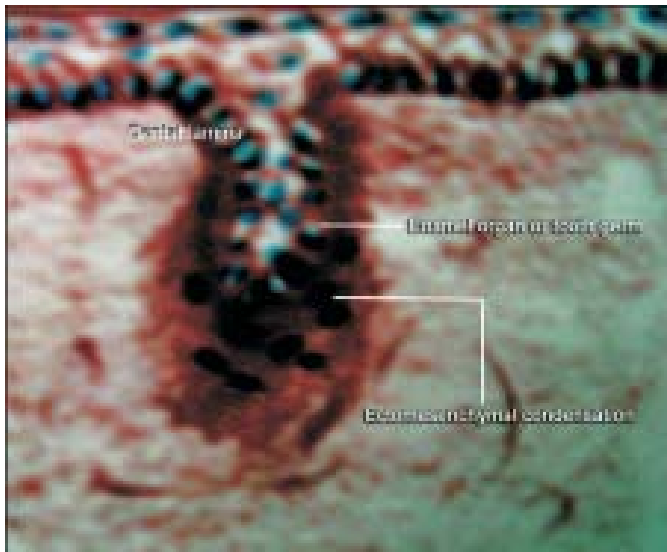


Fig. 2-2: Diagram of bud stage showing dental lamina and dental papilla formation (ectomesenchyme condensation).

Cap Stage (Proliferation)

- **Proliferation**—as the tooth bud continues to proliferate, there is unequal growth in different parts of the tooth bud, which leads to cap stage, which is characterized by shallow invagination on the deep surface of bud.
- **Enamel epithelium**—the peripheral cells of the cap, which cover the convexity is called as 'outer enamel epithelium' which is cuboidal cells and cells of concavity are called 'inner enamel epithelium' which is tall columnar cells (Fig. 2-3).
- **Stellate reticulum**—stellate reticulum is located between outer and inner enamel epithelium, which assume a reticular form. The space in this reticular network is filled with mucoïd fluid (rich in albumin) which gives it a 'cushion-like' consistency. Due to this, stellate reticulum supports and protects the delicate enamel-forming cells.



Fig. 2-3: Inner enamel epithelium cells further differentiate into ameloblasts. It also shows outer enamel epithelium (red arrow) and stellate reticulum (yellow arrow).

Bell Stage (Histodifferentiation and Morphodifferentiation)

- **Types of cell present**—as the invagination of epithelium deepens and its margins continue to grow; the enamel organ assumes 'a bell shape'. Four different types of cell i.e. cells of inner enamel epithelium, stratum intermedium, stellate reticulum and outer enamel epithelium are present (Fig. 2-4).
- **Cells of inner enamel epithelium**—it is single layer of tall columnar cells. The cells of inner enamel epithelium differentiate into ameloblast prior to amelogenesis. The cells of inner enamel epithelium exert an organizing influence on the underlying mesenchymal cells in dental papilla, which then differentiate into odontoblasts.
- **Stratum intermedium**—it is the squamous cells occurring between inner enamel epithelium and stellate reticulum.
- **Stellate reticulum**—these cells are star shaped. It has long process and it anastomoses with process of adjacent cells.
- **Outer enamel epithelium**—these are single layered cuboidal cells.
- **Enamel knot**—the cells in the center of the enamel organ are densely packed and form the enamel knot.
- **Enamel cord**—this is the vertical extension of the enamel knot. These cells are attached to one another by junctional complex laterally and to cells in stratum intermedium by desmosomes.
- **Membrana preformativa**—the basement membrane that separates the enamel organ and dental papilla just before dentin formation is called as 'membrana preformativa'.

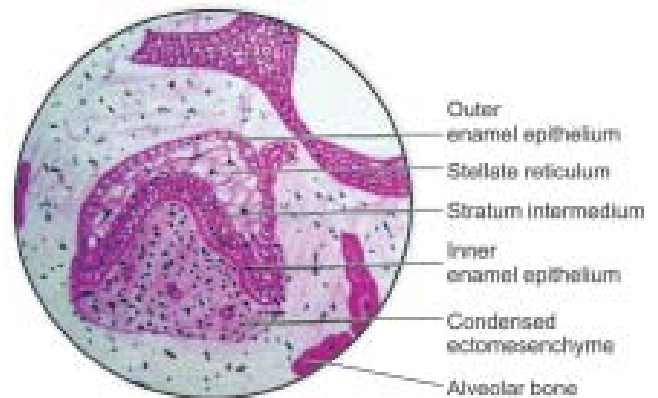


Fig. 2-4: Bell stage showing four different types of cells.

Advanced Bell Stage

- **Dentinoenamel junction**—during the advanced bell stage, the boundary between inner enamel epithelium and odontoblasts outline the future dentino-enamel junction.
- **Epithelial root sheath of Hertwig's**—in addition, cervical portion of enamel organ gives rise to epithelial root sheath of Hertwig's (Fig. 2-5).

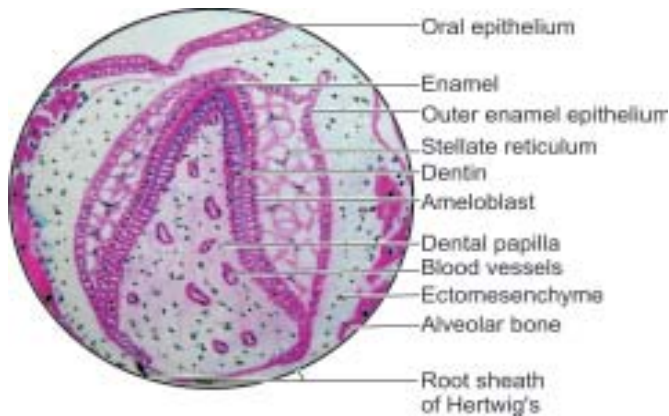


Fig. 2-5: Advanced bell stage showing root sheath of Hertwig's.

Eruption of Teeth

The axial or occlusal movement of tooth from its developmental position within the jaw to its functional position in the occlusal plane is known as *eruption of teeth*. There are 3 types of movements which are described as follows:

Pre-eruptive

- **Crowding of teeth**—when deciduous tooth germ first differentiates, there is good deal of space between them. But due to their rapid growth, this available space is utilized and developing teeth become crowded together, especially in incisor and canine region (Fig. 2-6).
- **Drifting of deciduous molar**—crowding is relieved by growth in length of infant's jaws, which provides room for second deciduous molars to drift backward and anterior teeth to drift forward. At the same time, the tooth germ also moves outward as jaw increases in width and height (Fig. 2-6).
- **Movement of permanent teeth**—permanent teeth with deciduous predecessors also undergo complete movement before they reach the position from which they will erupt (Fig. 2-6).
- **Eruption of deciduous predecessor teeth**—as their deciduous predecessors erupt, they move to a more apical position and occupy their own bony crypt.
- **Permanent premolars**—premolars begin their development lingual to their predecessors at the level of occlusal surface. They are situated beneath the divergent roots of deciduous molars.
- **Permanent molar**—the permanent molars which do not have predecessors also move from the site of their initial differentiation.

Eruptive

- **Axial movement**—there is axial or occlusal movement of tooth from its developmental position within the jaw to

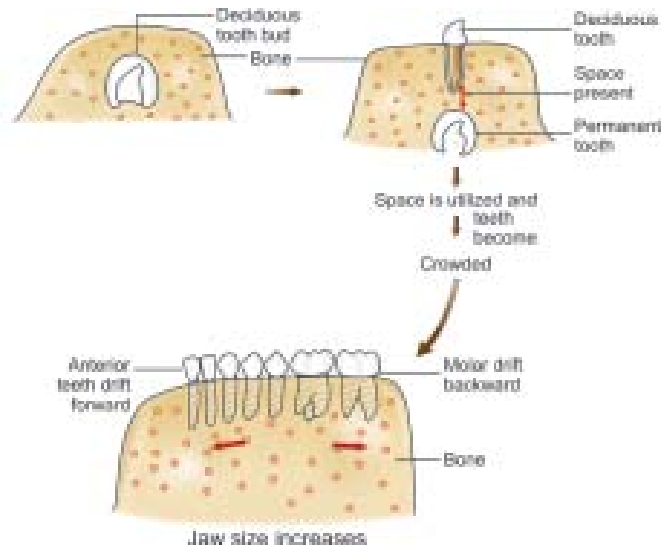


Fig. 2-6: Diagrammatic representation of pre-eruptive phase of eruption.

its final functional position in the occlusal plane. It is important to recognize that jaw growth is normally occurring while most of the teeth are erupting, so that movement in plane other than axial is superimposed on eruptive movement (Figs 2-7 and 2-8).

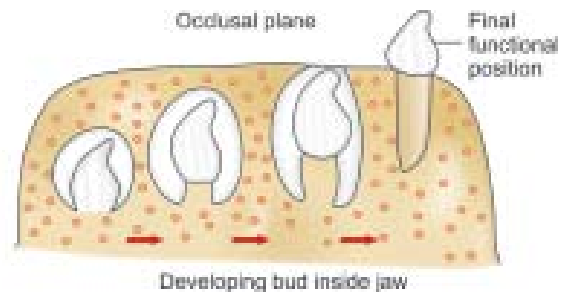


Fig. 2-7: Diagrammatic representation of eruptive phase of eruption.

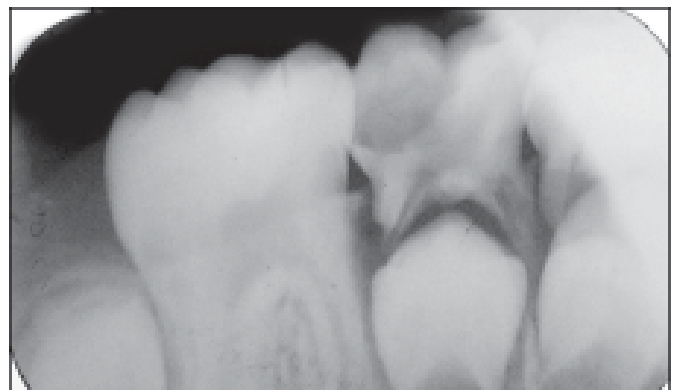


Fig. 2-8: Eruptive phase of eruption of teeth showing root resorption of deciduous molar.

Posteruptive

- **Maintenance**—it maintain the position of the erupted tooth while the jaw continued to grow.

Chronology of eruption of teeth				
Deciduous dentition				
Tooth	Formation of enamel matrix and dentin begins	Enamel completed	Eruption	Root completed
Upper				
Central incisor	4 months <i>in utero</i>	1½ months	7½ months	1½ years
Lateral incisor	4½ months <i>in utero</i>	2½ months	9 months	2 years
Cuspid	5 months <i>in utero</i>	9 months	18 months	3½ years
First molar	5 months <i>in utero</i>	6 months	14 months	2½ years
Second molar	6 months <i>in utero</i>	11 months	24 months	3 years
Lower				
Central incisor	4½ months <i>in utero</i>	2½ months	6 months	1½ years
Lateral incisor	4½ months <i>in utero</i>	3 months	7 months	1½ years
Cuspid	5 months <i>in utero</i>	9 months	16 months	3¼ years
First molar	5 months <i>in utero</i>	5½ months	12 months	2¼ years
Second molar	6 months <i>in utero</i>	10 months	20 months	3 years
Permanent dentition				
Tooth	First evidence of calcification	Crown completed	Eruption	Root completed
Upper				
Central incisor	3-4 months	4-5 years	7-8 years	10 years
Lateral incisor	10 months	4-5 years	8-9 years	11 years
Canine	3 years	6-7 years	11-12 years	13-15 years
First premolar	1½ - 1¾ years	5-6 years	10-11 years	12-13 years
Second premolar	2-2¼ years	6-7 years	10-12 years	12-14 years
1st molar	At birth	2½ - 3 years	6-7 years	9-10 years
2nd molar	2½ - 3 years	7-8 years	12-13 years	14-16 years
3rd molar	7-9 years	12-16 years	17-21 years	18-25 years
Lower				
Central incisor	3-4 months	4-5 years	6-7 years	9 years
Lateral incisor	3-4 months	4-5 years	7-8 years	10 years
Canine	4-5 months	6-7 years	9-10 years	12-14 years
1st pre molar	1¾ - 2 years	5-6 years	10-12 years	12-13 years
2nd pre molar	2¼ - 2½ years	6-7 years	10-12 years	13-14 years
1st molar	At birth	2½ - 3 years	6-7 years	9-10 years
2nd molar	2½ - 3 years	7-8 years	11-13 years	13-15 years
3rd molar	8-10 years	12-16 years	12-21 years	18-25 years

- *Compensatory growth*—it compensates for proximal and occlusal wear (Fig. 2-9).

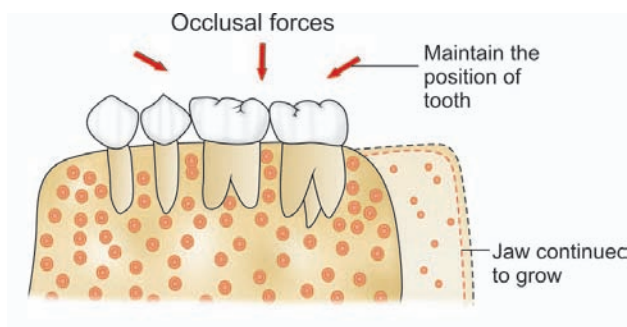


Fig. 2-9: Diagrammatic representation of post-eruptive phase of eruption.

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Development and Anatomy of Craniofacial Region

3

Oral Cavity Development

The primitive oral cavity is seen in third prenatal week. There is formation of pit in the tissue which underlies the forebrain. This pit is the future oral cavity. The formation of branchial arches occurs on either side of fetal neck, between the oral pit and developing heart. The process of development of oral cavity is as follows:

- *Invagination*—it occurs between forebrain and heart. Oral cavity forms under the forebrain.
- *Formation of oropharyngeal membrane*—this is wall formed between oral and pharyngeal cavity. It separates the stomodium from first part of foregut. Foregut will develop into pharynx.
- *Disintegration of oropharyngeal membrane*—in fourth week of intrauterine life, oropharyngeal membrane disintegrates. This will lead to continuity between oral and pharyngeal cavity.
- *Formation of endocrine gland*—endocrine glands can develop from roof and floor of oral cavity. Roof gives rise to Rathke's pouch, which results in the formation of anterior pituitary. Floor can give rise to second epithelial pouch which results in formation of endocrine tissues of the thyroid gland.
- *Formation of branchial arch*—tissues which surround the oral pit give rise to five to six pairs of branchial arch. The mandibular branchial arch is first arch to develop. The hyoid is second arch to develop. Other three arches are not so important in dental point of view.
- *Vestibule*—it is outer smaller portions of oral cavity. Vestibule of the mouth is a narrow space bounded externally by lips and cheeks and internally by teeth and gums.
- *Oral cavity proper*—it is inner larger part of oral cavity. It is bounded anterolaterally by the teeth, the gums and the alveolar arches of the jaws. The roof is formed by the hard and soft palate. The floor is occupied by the tongue posteriorly and sublingual region anteriorly, below the tip of tongue. Posteriorly, the cavity communicates with the pharynx through the oropharyngeal isthmus which is bounded superiorly by the soft palate, inferiorly by the tongue and on each side by the palatoglossal arches.
- *Arterial and venous supply of face*—arterial and venous supply is showing in **Figs 3-1 and 3-2**.

Vestibule

Lips

It is described in the Chapter 23: Disease of Lip.

Cheeks

- *Content*—cheeks are the fleshy flaps, forming a large part of the sides of the face. Mobile portion of cheeks is formed by the buccinator muscle. Intraorally, it is covered by the mucous membrane, and extraorally by the skin. The mucous membrane of the cheeks is fixed to the inner fascia of the buccinator muscle by tight strands of connective tissue.
- *Posterior part*—posterior part consist of masseter muscle and the parotid gland which are interposed between the mucous membrane and buccinator muscle on one side and the skin on the other side.
- *Nasolabial sulcus*—cheek are continuous in front with the lips and the junction is indicated by the *nasolabial sulcus* which extends from the side of nose to the angle of the mouth.

Anatomy of Oral Cavity

The oral cavity is incompletely bounded by bones. Its lateral and anterior walls are formed by the inner surface of the alveolar processes, which join at the midline. The lingual surface of the teeth completes these walls. Oral cavity is divided into:

- *Buccal fat pad of Bichet*—the cheek contains a peculiar body of fat tissue called as *buccal fat pad of Bichet*. It is rounded biconvex structure limited by a thin but distinctive capsule.
- *Blood supply*—it is supplied by the branches of the maxillary artery (Fig. 3-1).
- *Lymphatic drainage*—drain into the submandibular and pre-auricular lymph nodes and partly to the buccal and mandibular nodes.

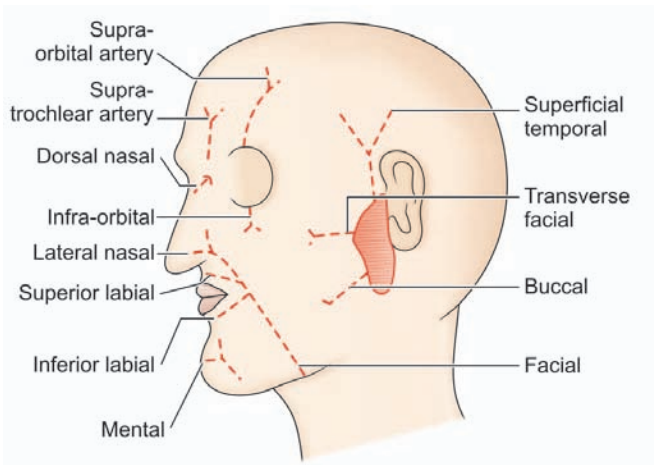


Fig. 3-1: Arterial supply of the face.

Oral Cavity Proper

Gingiva

It is described in the Chapter 24: Gingival and Periodontal Diseases.

Teeth

- *Structure*—the teeth form a part of the masticatory apparatus and are fixed to the jaws. In man, the teeth are replaced only once (*diphyodont*) in contrast with non-mammalian vertebrates where teeth are constantly replaced throughout life (*polyphyodont*). Each tooth has three parts, i.e. crown (projection above the gums), root (embedded in the jaw beneath the gum) and neck (between the crown and root and surrounded by the gums).
- *Nerve supply*—it is supplied by *anterior superior alveolar* (upper incisor and canine teeth), *middle superior alveolar* (upper premolar teeth), *posterior superior alveolar* (molar teeth) and *inferior alveolar nerve* (lower teeth).
- *Blood supply*—it is supplied by *posterior superior alveolar artery* (molar and premolar maxillary teeth), *anterior superior alveolar* (It is branch of infraorbital artery and supplies incisor and canine maxillary teeth) and *inferior alveolar artery* (it enters the mandibular canal and gives branches to the mandible and to the roots of each teeth attached to the bone).

Hard Palate

- *Development*—this is the tissue which is interposed between oral and nasal cavity. Palate develops from medial and lateral palatine process. Development of palate starts in sixth week. It develops as intermaxillary segment, between maxillary process of upper jaw. This is called as primary palate. At the end of sixth week, secondary palate develops from lateral palatine process. Lateral palatine process grows medially downward or vertically on either side of tongue.
- *Boundaries*—it is a partition between the nasal and oral cavities. Anterolateral margins are limited by alveolar arches and gingiva. Posterior margin is continuous with the soft palate. Superior surface forms the floor of the nose and inferior surface forms the roof of the oral cavity.
- *Nerve supply*—it is supplied by greater palatine nerves from the greater palatine foramen and nasopalatine nerve from the incisive foramen.
- *Blood supply*—it is supplied by greater palatine branch of the maxillary artery and nasopalatine artery.
- *Venous drainage*—palatine vessels go to the pterygoid plexus of veins.
- *Lymphatic drainage*—it drains mostly into the upper cervical and partly into the retropharyngeal groups of nodes.

Soft Palate

- *Content*—it is a movable fold suspended from the posterior border of the hard palate. It separates the nasopharynx from the oropharynx. It has two surfaces, i.e. anterior and posterior and two borders, i.e. superior and inferior.
- *Anterior surface*—it is concave and is marked by median raphe.
- *Posterior surface*—it is convex and is continuous superiorly with the floor of the nasal cavity.
- *Superior border*—it is attached to the posterior border of the hard palate, blending on each side with pharynx.
- *Inferior border*—it is free and bounds with pharyngeal isthmus.
- *Muscle of the soft palate*—these are tensor palati, levator palati, musculus uvula, palatoglossal and palatopharyngeus.
- *Nerve supply*—all muscle of the soft palate except the tensor palati are supplied by the pharyngeal plexus. The fibers of the plexus are derived from the cranial part of the accessory nerve. The tensor palati is supplied by mandibular nerve. General sensory nerves are derived from lesser palatine nerve.
- *Blood supply*—greater palatine branch of the maxillary artery, ascending palatine branch of facial and palatine branch of ascending pharyngeal arteries.

- *Venous drainage*—veins pass to the pterygoid and tonsillar plexus of veins.
- *Lymphatic drainage*—lymphatics drain into upper cervical and retropharyngeal lymph nodes.

Tongue

It is described in the Chapter 22: Diseases of Tongue.

Floor of Mouth

- *Content*—it is a crescent shaped area between the lower gingiva and undersurface of the tongue which composes the inferior most portion of the oral cavity overlying the mylohyoid and thyroglossal muscles.
- *Nerve supply*—it is supplied by the branches of trigeminal nerve.
- *Arterial supply*—it is supplied by facial artery.
- *Venous drainage*—drains into facial or lingual vein (**Fig. 3-2**).
- *Lymphatic drainage*—from the anterior portion of mouth, lymphatics may pass into the deep cervical nodes or laterally to the periosteal lymphatics and then to the submandibular nodes and goes to the deep internal jugular nodes.

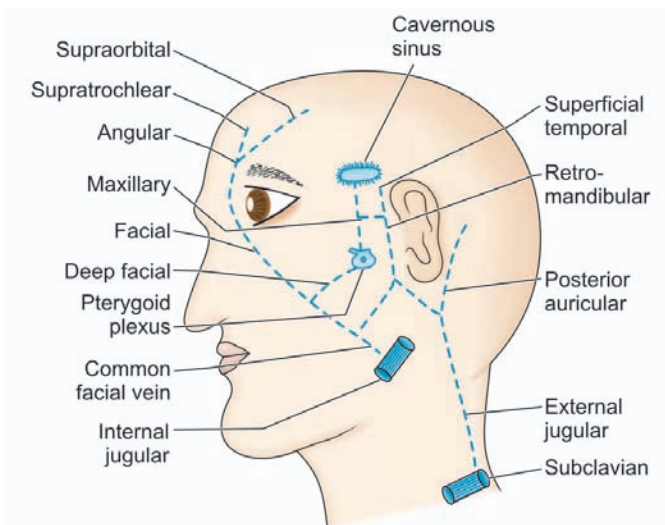


Fig. 3-2: Venous supply of the face.

Maxillary Sinus

It is described in the Chapter 27: Disorders of Maxillary Sinus.

Salivary Glands

It is described in the Chapter 26: Disorders of Salivary Glands.

Pharynx

- *Development*—muscles of pharynx are formed at about 7th week of intrauterine life. It forms from the muscle

cell of third and fourth arches. It forms the stylopharyngeal, cricothyroid, levator palatine and constrictor muscle of the pharynx.

- *Content*—it is a wide muscular tube situated behind the nose, mouth and larynx. Clinically, it is a part of upper respiratory tract. It is divided into three parts, i.e. nasopharynx (nasal part of pharynx), laryngopharynx (laryngeal part of pharynx) and oropharynx (oral part of pharynx). Oropharynx is the middle part of the pharynx situated behind the oral cavity.
- *Blood supply*—it is supplied by ascending pharyngeal branch of the external carotid artery, ascending palatine and tonsillar branch of the facial artery, dorsal lingual branch of lingual artery and greater palatine, pharyngeal and pterygoid branch of the maxillary artery.
- *Venous drainage*—it is supplied by a plexus which receives blood from the pharynx and soft palate and prevertebral region and drains into the internal jugular and facial veins.
- *Lymphatic drainage*—it drains into the retropharyngeal and deep cervical lymph nodes.
- *Nerve supply*—it is supplied by the pharyngeal plexus of nerves which lies chiefly on the middle constrictor.

Muscles of Mastication

The muscles of mastication move the mandible during mastication and speech. They are the masseter, the temporalis, the lateral pterygoid and the medial pterygoid.

Development

- *Proliferation of myoblasts*—muscles of mastication are derived from mandibular arch, i.e. first branchial arch. In fifth and sixth week of intrauterine life, proliferation of myoblasts occurs.
- *Orientation of muscle cells*—muscle cells become oriented to the sites of origin and insertion.
- *Migration*—enlargement of muscle mass occurs and it will migrate into the areas of differentiation.
- *Formation of muscles*—after this, it will be differentiated into masseter, medial and lateral pterygoid and temporal muscle. By tenth prenatal week, muscle mass becomes well organized. Muscle cells of masseter and medial pterygoid form vertical lob which is inserted at angle of mandible. Fibers of lateral pterygoid go horizontally and insert in the articular disc. The temporalis muscle has differentiated in the infratemporal fossa and is inserted in the coronoid process (**Fig. 3-3**).
- *Innervations of facial muscle*—in seventh week, fifth nerve enters the mandibular arch and seventh nerve in second branchial arch.

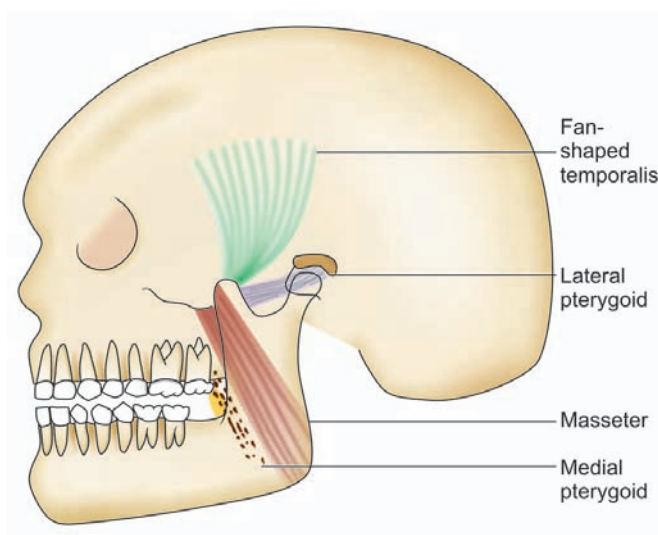


Fig. 3-3: Muscle of mastication—A diagrammatic representation.

Masseter Muscle

- **Site**—it is the most superficial to the masticatory muscle, stretches as a rectangular plate from the zygomatic arch to the outer surface of the mandible. It has three layers i.e. superficial, middle and deep.
- **Superficial layer**—it arises by thick aponeurosis from the zygomatic process of the maxilla and from the anterior two-third of the lower border of zygomatic arch. Its fibers pass downward and backward to be inserted into the angle and lower half of lateral surface of ramus of mandible (Fig. 3-4).
- **Middle layer**—it arises from the deep surface of the anterior two-third of the zygomatic arch and posterior one-third of lower border of zygomatic arch and is inserted into middle of ramus of mandible.
- **Deep layer**—it arises from deep surface of the zygomatic arch and is inserted into upper part of the ramus of the mandible and into the coronoid process.
- **Nerve supply**—it is supplied by masseteric nerve which is a branch of anterior division of the mandibular nerve.
- **Blood supply**—the masseteric artery which is a branch of internal maxillary artery and the masseteric vein follow the course of the nerve.

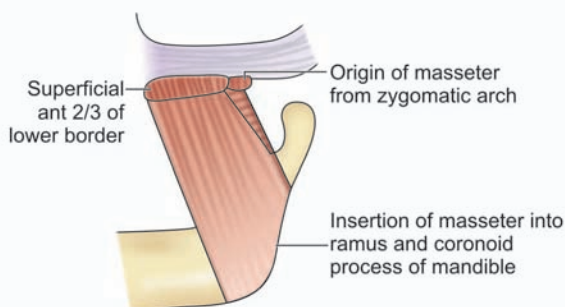


Fig. 3-4: Origin and insertion of masseter muscle.

- **Functions**—its main function is elevation of mandible, its superficial layer may also aid in protruding the mandible. When the mandible is protruded and biting force is applied, the fibers of the deep portion stabilize the condyle against the articular eminence.

The Temporalis Muscle

- **Origin and insertion**—it is fan shaped and arises from whole of the temporal fossa and from the deep surface of temporal fascia. Its fibers converge and descend into tendon which passes through the gap between the zygomatic arch and the side of the skull to be attached to the medial surface, apex, anterior and posterior borders of the coronoid process and the anterior border of the ramus of mandible nearly as far as the last molar teeth (Fig. 3-5).
- **Nerve supply**—it is supplied by the two deep temporal branches of anterior trunk of the mandibular nerve.
- **Blood supply**—it is supplied by middle and deep temporal arteries. The middle temporal artery is a branch of the superficial temporal artery. The deep temporal artery is a branch of internal maxillary artery.
- **Function**
 - When the entire temporalis contract, it elevates the mandible.
 - Its middle fibers have a retracting component because of their oblique direction downward and forward.
 - Its posterior fibers retract the protruded mandible.

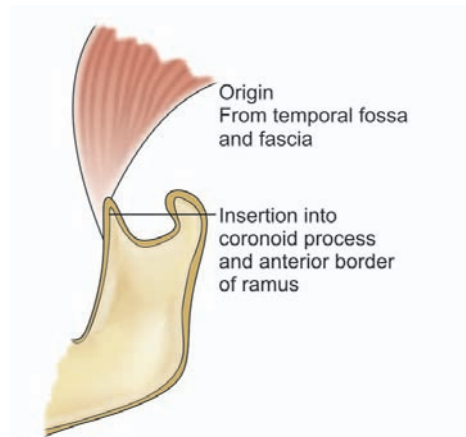


Fig. 3-5: Origin and insertion of temporalis muscle.

Lateral Pterygoid

- **Origin and insertion**—It is a short thick muscle with two heads. Upper arises from the infratemporal surface and infratemporal crest of the greater wing of sphenoid bone and lower from the lateral surface of lateral pterygoid plate. Its fibers pass backward and laterally to be inserted into the pterygoid fovea on the anterior surface of the neck of the mandible and into the articular capsule and disc of temporomandibular joint (Fig. 3-6).

- **Nerve supply**—it is supplied by a branch of the anterior trunk of mandibular nerve.
- **Blood supply**—branch of maxillary artery.
- **Function**
 - It assists in opening the mouth by pulling forward the condylar process of the mandible and the articular disc, while the head of the mandible rotates on the articular disc.
 - During closure of mouth, backward gliding of the articular disc and condyle of the mandible are controlled by slow elongation of lateral pterygoid with medial pterygoid of the same side.
 - The medial and lateral pterygoid muscle of both sides contract alternately to produce side-to-side movement of the mandible.
 - When medial and lateral pterygoid of both sides act together, they protrude the mandible.

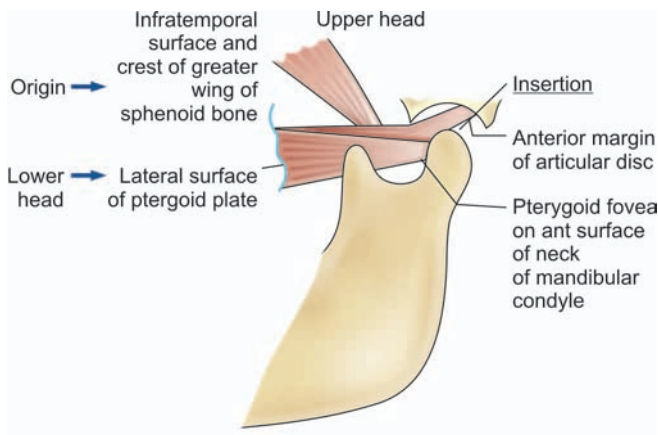


Fig. 3-6: Origin and insertion of lateral pterygoid muscle.

Medial Pterygoid

- **Origin and insertion**—it is a thick quadrilateral muscle attached to the medial surface of lateral pterygoid plate and the grooved surface of the pyramidal process of the palatine bone above. It has a superficial head which originates from the tuberosity of the maxilla and adjoining bone. Its deep head originates from the medial surface of medial pterygoid plate and the lateral surfaces of pyramidal process of palatine bone. Its fibers pass downward, laterally and backward and are attached by strong tendinous lamina to the posterior inferior part of the medial surfaces of the ramus and the angle of mandible as high as mandibular foramen and as forward as mylohyoid groove (Fig. 3-7).
- **Nerve supply**—it is supplied by branch of the mandibular nerve.
- **Blood supply**—it is supplied by the branch of maxillary artery.
- **Functions**—it helps in the elevation of mandible. Acting with the lateral pterygoid, they protrude the mandible.

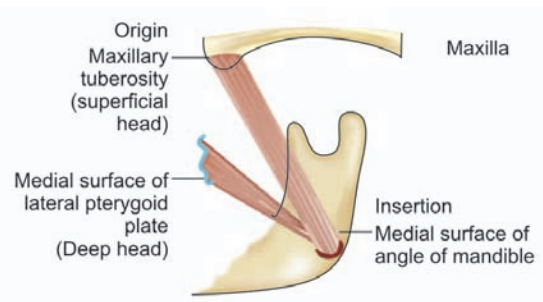


Fig. 3-7: Origin and insertion of medial pterygoid muscle.

Bones

The skull consists of the 22 bones. Out of which, 8 are paired and 6 are unpaired. Paired bones are parietal, temporal, maxilla, zygomatic, nasal, lacrimal, palatine and inferior nasal concha. Unpaired bones are frontal, occipital, sphenoid, ethmoid, mandible and vomer. From dental point of view, maxilla and mandible are the most important and they are described below:

Maxilla

The maxilla consists of a central body, which is hollowed out forming the maxillary sinus and four processes (Fig. 3-8).

- **Frontal process**—it ascends from the anteromedial corner of the body, serves as the connection with the frontal bone.
- **Zygomatic process**—it forms in the lateral corner of the body, connects with the zygomatic bone.
- **Palatine process**—it is horizontal and arises from the lower edge of the medial surface of the body.
- **Alveolar process**—it extends downwards and carries the socket for the maxillary teeth.

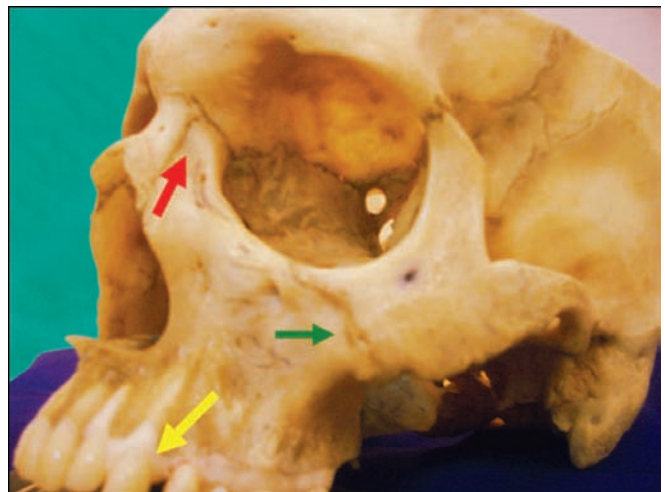


Fig. 3-8: Maxilla showing frontal process (red arrow) zygomatic process (green arrow) and alveolar process (yellow arrow).

- *Body of maxilla*—the body of maxilla is three side pyramid with its base facing the nasal cavity (**Fig. 3-9**). It lies in an almost horizontal axis with its apex being elongated into the zygomatic process.

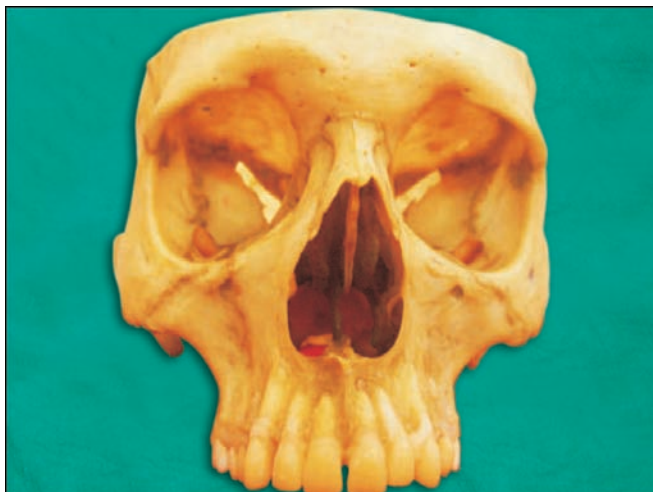


Fig. 3-9: Front view of maxilla.

- *Side of maxilla*—the three sides are superior or orbital (it forms greater part of the orbital floor), an anterolateral or malar (surface forming part of the skeleton of the cheek and face) and posterolateral or infra-temporal (surface turned towards the infra-temporal fossa).
- *Base*—the base is rimmed on its inferior edge by the alveolar process housing the teeth row.
- *Alveolar process*—alveolar process consists of two roughly parallel plates of bone that unite behind the last tooth to form a small rough prominence, the alveolar



Fig. 3-10: Palatal view of maxilla showing incisive foramen (red arrow).

tubercle, which often contains a single large marrow process. The lateral and external alveolar plate continues upward into the anterolateral and posterolateral surface of the maxillary body. The internal alveolar plate continues into the palatine process and behind the posterior end of the latter into the nasal surface of the maxillary body. The deep furrow between the two alveolar plates is divided by radial bony plates into the sockets of the individual teeth.

- *Incisive foramina*—at the boundary between two portions of the nasal crest, a canal commences in the nasal floor close to the midline and extends downwards, anteriorly and medially to unite with the canal of the other side in a common opening which is called as *incisive or nasopalatine canal* (**Fig. 3-10**). On the anterior surface of maxilla, there is canine fossa situated lateral to the canine eminence.

Mandible

It is the largest and strongest bone of the face. It consists of a horseshoe shaped body continuous upward and backward on either side with the mandibular rami.

- *Body*—the body is thick, has a rounded lower border and carries the alveolar process on its upper border. It extends backward from the chin at the midline symphysis to the anterior limit of the ramus.
- *Ramus*—it is a thick quadrilateral plate which extends backward from the groove for the facial artery (antegonial notch) to include the region called the mandibular angle (**Fig. 3-11**). The anterior border of the ramus continues along the body lateral to the alveolar process as a blunt ridge, the oblique line, running downward and forward to disappear at about the level of the 1st molar.



Fig. 3-11: Overview of mandible showing body and ramus of mandible.

- *Mental protuberance*—in the midline of anterior surface of the body projects a triangular prominence called mental protuberance.
- *Symphysis menti*—it is the line at which the right and left halves of the bone meet each other.
- *The mental foramen*, through which the mental nerve and blood vessels pass, is located on the lateral surface of body between the roots of the 1st and 2nd premolars. In the vertical dimension, the foramen lies halfway between the lower border of mandible and the alveolar margin.
- *Genial tubercle*—slightly above the lower border on its inner surface the mandibular symphysis is elevated in more or less sharply defined projection called as genial tubercle.
- *Mylohyoid line*—it is a prominent ridge that runs obliquely downward and forward from below 3rd molar tooth to the medial area below the genial tubercle. Below the mylohyoid line, the surface is slightly hollowed out to form submandibular fossa, which lodges the submandibular gland.
- *Mandibular canal*—the mandibular canal which houses the inferior alveolar nerve and blood vessels begins at the mandibular foramen, curves downward and forward and turns into a horizontal course below the roots of the molars. In the region of the premolars, the mandibular canal splits into two canals of unequal width; the narrower incisive canal continues the course of mandibular canal toward the midline and the wider branch, the mental, turns laterally, superiorly and posteriorly to open at the mental foramen (Fig. 3-12).

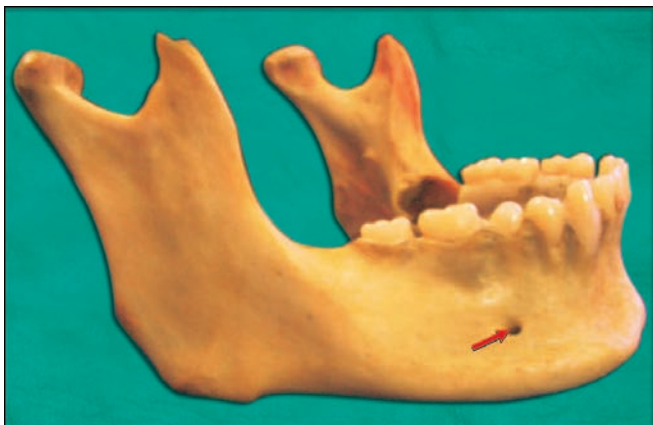


Fig. 3-12: Side view of mandible showing mental foramina (red arrow).

- *Alveolar bone*—the alveolar plates consist of two compact bony plates, the external and internal alveolar plate. These two plates are joined to each other by radial interdental and in the molar region, by inter-radicular septa, thus forming the sockets for the teeth in the same manner as in the upper jaw.

- *Digastric fossa*—the lower border of the mandible is also called as base. Near the midline, the base shows an oval depression called *digastric fossa*.

Trigeminal Nerve

The trigeminal nerve is the 5th cranial nerve and is also the largest. It has a large sensory root and a small motor root. It is attached to lateral part of pons by its two roots. It conveys both exteroceptive and proprioceptive impulses. Exteroceptive impulses of touch, pain and thermal senses are transmitted from skin of face, forehead, mucous membrane of nasal and oral cavity, sinus, and floor of mouth, teeth and anterior 2/3rd of tongue. Proprioceptive impulses of deep pressure are conveyed from teeth, periodontium, hard palate and temporomandibular joint receptors.

Branches of Trigeminal Nerve (Fig. 3-13)

Ophthalmic division

It is the smallest branch of semilunar ganglion and passes forward in the lateral wall of cavernous sinus.

- *Lacrimonal nerve*—it supplies sensory fibers to the gland and the adjacent conjunctiva.
- *Frontal nerve*—it divides into *supraorbital nerve* (supplies the skin of the upper eyelid, forehead and the anterior scalp region to the vertex of skull), and *supratrochlear nerve* (supplies skin of the upper eyelid and lower medial portion of forehead).
- *Nasociliary nerve*—it gives numerous branches. It includes branches in orbit (*Long ciliary nerves, posterior ethmoid nerve, anterior ethmoid nerve, external nasal branches*), branches arising in nasal cavity and terminal branches on the face.

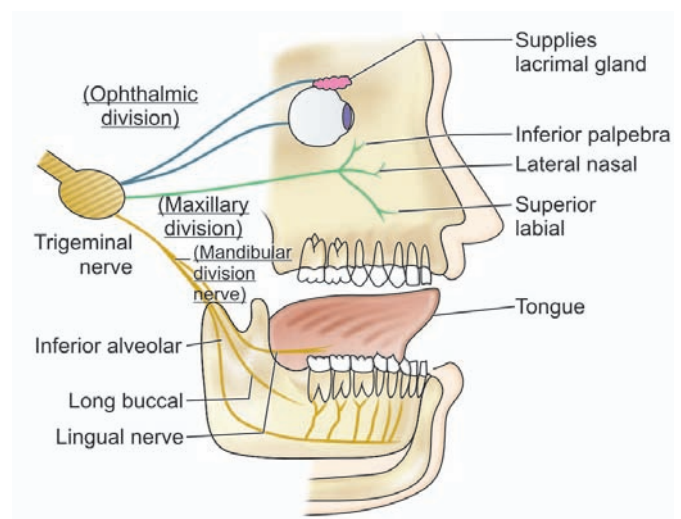


Fig. 3-13: Different branches of trigeminal nerve supplying to face.

Maxillary division

It is intermediate division, and entirely sensory. It enters the orbit through inferior orbital fissure. It is now named *infraorbital nerve* and having traversed the infraorbital groove and canal in floor of orbit, it appears on face through infraorbital foramen. Branches are divided into 4 groups

- *Middle meningeal nerve*—supplies the dura with sensory fibers.
- *Ganglionic branches*—they contain secretomotor fibers for the lacrimal gland and sensory fibers for orbital periosteum and mucous membranes of the nose, palate and pharynx.
- *Zygomatic nerve*—it is divided into two branches, i.e. *zygomaticofacial* (supplies sensory fiber to skin over the prominence of zygomatic bone) and *zygomaticotemporal* (it supplies sensory fibers to skin over anterior temporal fossa region).
- *Posterior superior alveolar nerve*—it gives sensory branches to mucous membrane of sinus. It also supplies the maxillary molars and their gingivae.
- *Middle superior alveolar nerve*—it supplies upper premolar and mesiobuccal root of upper first molar.
- *Anterior superior alveolar nerve*—it supplies roots of maxillary central and lateral incisors. They also send branches to superior dental plexus of nerves within maxilla. They also supply mucous membrane of anterior part of maxillary sinus as well as labial gingivae of incisors and cuspid teeth.
- *Inferior palpebral branches*—they supply sensory fibers to skin of lower eyelid and its conjunctiva.
- *External or lateral nasal branches*—they supply skin of side of nose.

- *Superior labial branches* - three or more in number and supply skin and mucous membrane of upper lip and labial glands.

Mandibular Nerve

It is the largest of the three divisions. It is divided into following branches:

- *Nervus spinosus*—it supplies dura and mastoid cells.
- *Nerve to internal pterygoid muscle*—it supplies internal pterygoid muscle.
- *Pterygoid nerve*—it enters medial side of external pterygoid muscle to provide motor nerve supply.
- *Masseter nerve*—it supplies masseter muscle.
- *Anterior deep temporal nerve*—it ends in deep part of anterior portion of temporal muscle.
- *Posterior deep temporal nerve*—it passes upward to deep part of temporal muscle.
- *Long buccal nerve*—it supplies mandibular 2nd and 3rd molars. It then sends fibers to mucous membrane and skin of cheek, retromolar triangle, and buccal gingivae of mandibular molars and mucous membrane of lower part of buccal vestibule.
- *Auriculotemporal nerve*—it traverses upper deep part of parotid gland and then crosses the posterior root of zygomatic arch. It passes with superficial temporal artery in its upward course and then divides with numerous branches to tragus of external ear, to scalp, to the ear and as far upward as vertex of skull.
- *Parotid branches*—they are sensory, secretory and vasomotor fibers to the gland.
- *Articular branches*—it enter the posterior part of the temporomandibular joint.

TABLE 3-1: Lymphatic drainage of head and neck

<i>Nodes</i>	<i>Sites</i>
Occipital	Scalp, posterior to the ear and occipital region
Posterior auricular	External ear, scalp above and behind the ear.
Anterior auricular (pre-auricular/parotid)	Skin anterior to the temple, external meatus, lateral forehead, lateral eyelids, infraorbital nodes, posterior cheek, part of the outer ear, parotid gland
Inferior auricular (infra-auricular)	Pre- and post-auricular nodes
Infra-orbital	Skin of inner corner of the eye, skin of anterior face, and superficial aspect of the nose
Buccal	Skin over the anterior face, mucous membrane of the lips and cheeks, occasionally mandibular and maxillary teeth and gingivae
Submental	Tip of the tongue, midportion of the lower lips, chin, lower incisors and gingivae
Mandibular (supra-mandibular)	Skin over the mandible, mucous membrane of the lips and cheeks. Occasionally, maxillary and mandibular teeth and gingivae.
Submandibular (sub-maxillary)	Upper and lower teeth and gingivae except mandibular incisor, anterior nasal cavity, palate, body of tongue, upper lip, lateral angle of eye, submental nodes
Superficial cervical	Pinna and adjacent skin, pre- and post-auricular nodes
Deep cervical	Submandibular, submental, inferior auricular, tonsillar and tongue nodes

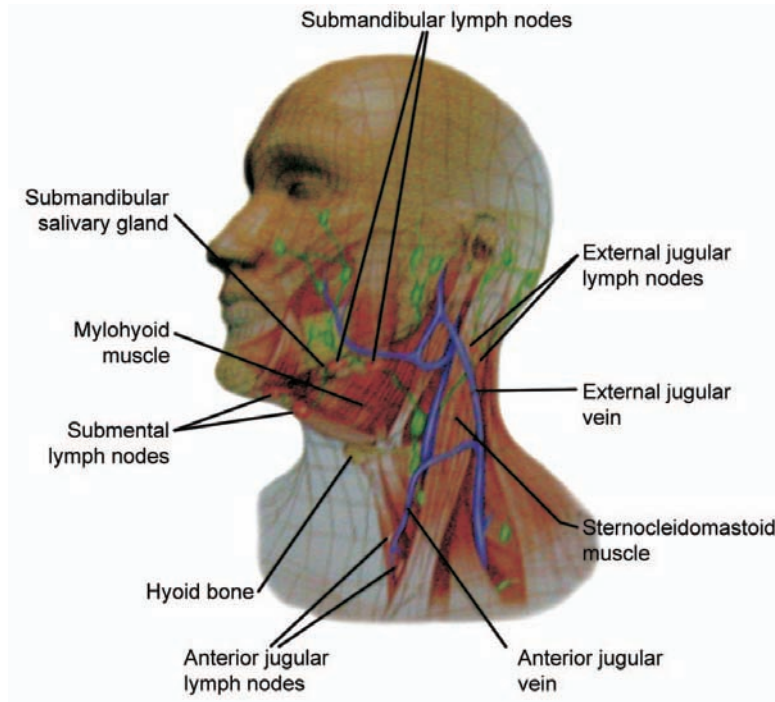


Fig. 3-14: Lymphatic drainage of face.

- *Auricular branches*—it supplies the skin of helix and tragus.
- *Meatal branches*—two small branches which supply skin lining the meatus and tympanic membrane.
- *Terminal branches*—they supply scalp and temporal region.
- *Lingual nerve*—It gives off small branches that are sensory to part of tonsil and mucous membrane of posterior part of oral cavity. It is sensory to mucous membrane of oral cavity, anterior 2/3rd of tongue (along with chorda tympani nerve), floor of mouth and gingivae on lingual surface of the mandible.
- *Inferior alveolar nerve*—it is the largest branch of posterior division of mandibular nerve. It sends motor branches to mylohyoid muscle and anterior belly of digastric muscle. It then enters mandibular foramen and descends in the mandible in the inferior dental canal as inferior alveolar nerve. It is sensory to mandibular teeth, body of mandible and labial gingiva anterior to bicuspid teeth.
- *Mental nerve*—it passes through mental foramen on lateral surface of mandible. It is sensory to skin of chin, lower lip, and mucous membrane lining of lower lip.
- *Incisive nerve*—it continues anteriorly in the inferior dental canal to midline. It is sensory to anterior teeth and labial gingivae.

Lymphatic Drainage of Head and Neck

It is described in **Table 3-1** and **Fig. 3-14**.

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Immunity, Antigen-Antibody Reaction

Immunity

The word immunology derived from Latin word 'immunis' meaning 'free of burden'. "It is the resistance exhibited by the host towards injury caused by microorganisms and their products". It is a reaction of body against any foreign antigens. Immunity against infectious diseases consists of two main types, each with humoral and cellular components and their effective cells. The importance of immune system occurs in life-threatening infection suffer by patient with immune defect.

Uses of Immunity

- *Understanding the disease*—it helps to understand the etiology and pathogenesis of many diseases.
- *Vaccine*—development of vaccine can be done with the help of immunity.
- *Treatment*—treatment of many diseases can be done with antibodies.
- *Future susceptibility*—it helps to find with future susceptibility to disease with the help of HLA typing system.

Classification (Fig. 4-1)

- **Innate immunity**
 - Non-specific
 - Specific
- **Acquired**
 - Active
 - Natural
 - Artificial
 - Passive
 - Natural
 - Artificial
- **Local**
- **Herd**

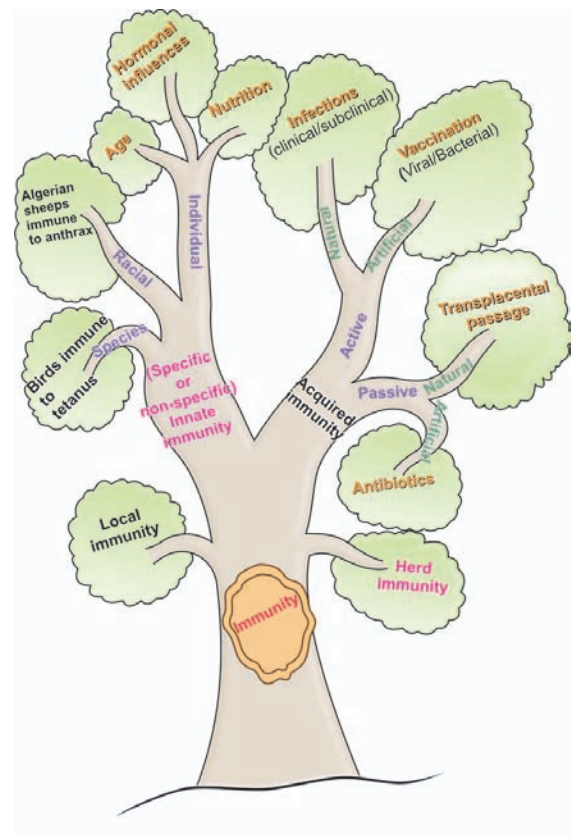


Fig. 4-1: Different types of immunity—
A diagrammatic representation.

Innate Immunity

This is also called as natural immunity. This comprises of preexisting non-specific defences. It is the resistance to infection, which an individual possesses by virtue of his genetic and constitutional make up. It does not depend upon the prior contact with microorganisms or immunization. Innate immunity can be considered at the level of race, species or at individual's levels.

Types of Innate Immunity

- *Specific and non-specific*—it may be *non-specific* when it indicates degree of resistance to infection in general or *specific* when resistance to particular pathogen is concerned.
- *Species immunity*—it refers to total or relative refractoriness to a pathogen shown by all members of a species e.g. all human beings are totally unsusceptible to plant pathogens and to many pathogens of animals, such as rinderpest or distemper. It may be due to physiological and biochemical differences between the tissues of different host species, which determine whether a pathogen can multiply or not.
- *Racial immunity*—within species, different races may show differences in susceptibility to infection. This is called as 'racial immunity' e.g. high resistance of Algerian sheep to anthrax. Such racial differences are known to be genetic in origin.
- *Individual immunity*—the differences in individual immunity exhibited by different individuals in a race is called as 'individual immunity' e.g. homozygous twins exhibit similar degree of resistance or susceptibility to lepromatous leprosy and tuberculosis. Such correlation is not seen in heterozygous twins.

Factors Affecting Innate Immunity

- *Age*—the two extremes of life carry higher susceptibility to infection as compared to adults. The fetus *in utero* however is protected from maternal infection by the placental barrier.
- *Hormonal influence*—endocrine disorders such as diabetes mellitus, hypothyroidism and adrenal dysfunction are associated with an enhanced susceptibility to infection. Corticosteroids exert an important influence on response to infection. The elevated steroid levels during pregnancy may have a relation to heightened susceptibility of pregnant women to many infections.
- *Nutrition*—in general, both humoral and cell mediated responses are reduced in malnutrition. Cell mediated immune response such as *Mantoux* test becomes negative in severe protein deficiency. Certain infections may not become clinically apparent in severely malnourished patients.

Mechanism of Innate Immunity

- *Epithelial surface*—skin and mucous membrane covering the body gives protection against bacteria. They act as mechanical barrier.
- *Humoral factors*—it consists of lysozyme, properdin, betalysin, C-reactive protein, bactericidin etc.
- *Cellular factors*—it includes phagocytosis, and inflammation.

Acquired Immunity

The resistance that an individual acquires during life is known as '*acquired immunity*'. It is of two types.

Active Immunity

It is resistance developed by an individual because of antigenic stimulus. This involves active functioning of host's immune apparatus leading to synthesis of antibodies or production of immunologically active cells. Once the active immunity develops, it is long lasting. It is also of two types.

- *Natural*—it results from either a clinical or an inapparent infection with the parasite, e.g. a person who has recovered from an attack of smallpox develops natural immunity to it. The immunity following bacterial infection is generally less permanent than that following a viral infection.
- *Artificial*—it is the resistance induced by vaccines, which are preparations of live or killed microorganisms or their product.

Passive Immunity

The resistance that is transmitted to a person in readymade fashion is known as passive immunity. There is no antigenic stimulant, instead preformed antibodies are administered. There is no latent period, protection being effective immediately after passive immunization. It is less effective and inferior to active immunization, but it is immediate in action and can be employed when instant immunity is needed. It is also of two types.

- *Natural*—it is the resistance passively transferred from mother to baby. By active immunization of mothers during pregnancy, it is possible to improve the quality of passive immunity in the infants.
- *Artificial*—it is passively transferred to the recipient by the administration of antibodies. The agent used is hyperimmune sera of animal or human origin, convalescent sera and pooled human gamma globulin.

Local Immunity

It is important in treatment of infection, which is either localized or due to surgeries (postoperative infection), in combating infections at the site of primary entry of the pathogen. Natural infection or the live virus vaccine administered provides local immunity.

Herd Immunity

This refers to the overall level of immunity in a community and is relevant in the control of epidemic diseases. When a large proportion of individuals in a community (herd)

is immune to a pathogen, herd immunity to a pathogen is satisfactory. When herd immunity is low, epidemics are likely to occur on the introduction of a suitable pathogen.

Antigen-Antibody Reaction

Antigen

- **Definition**—any substance which when introduced parenterally into a body stimulates the production of an antibody, with which it reacts specifically and in an observable manner. The immune system can respond to antigen either by cell mediated immunity or by humoral immunity.
- **Size**—most antigens are large molecules (over 1000 molecular weight). Smaller molecules do not provoke an immune response unless bound to large carrier molecules. The complete antigen is able to induce antibody formation and produces a specific and observable reaction with the antibody so produced.
- **Haptens**—these are substances which are incapable of inducing antibody formation by themselves, but can react specifically with antibodies.
- **Epitope**—the smallest unit to antigenicity is known as an *epitope* or antigenic determinant.

Antibody

Antibody is produced by plasma cells in the lymph nodes, bone marrow and spleen. The cells are ovoid with an eccentrically placed nucleus. The cytoplasm is basophilic. One plasma cell produces antibody of one class, reactive with only one antigen. There are five classes of immunoglobulins which are as follows:

IgG

It is the most abundant immunoglobulin in the plasma and extracellular fluid. It can cross placenta and is important in passive transfer of immunity to the fetus. It is capable of neutralizing toxins and may be cytolytic through the activation of a complement. Polymorphs and macrophages have surface receptors for Fc fragment of IgG, thus binding of IgG to particular antigens promotes adhesion of these cells and subsequent phagocytosis of antigen.

IgA

It is secreted locally by plasma cells in the respiratory passages, salivary and lacrimal glands and intestinal mucosa. It is an important constituent of breast milk. IgA occurs in two forms, serum IgA is principally a monomeric

7S molecule found on mucosal surfaces and in secretions. It is a dimer formed by two monomer units joined together at their carboxyterminals by glycopeptides termed the 'J chain'. This is called as secretory IgA. It can activate complement by the alternative pathway.

IgM

It is formed by J chains into pentamers of the Ig molecules and these attain very high molecular weight of 9000,000. The large molecular size prevents it from leaving the plasma, except when permitted by increased vascular permeability in inflammatory lesions. As it has antigen combining sites, it has good agglutinating and complement fixing properties. It is the first class of antibodies to be formed in immune response.

IgE

It binds selectively to mast cells and to basophils by its Fc fragment. The binding of antigen to its Fab fragment triggers reflex of histamine and other substances which are important in anaphylactic type of hypersensitivity.

IgD

The function of IgD is largely unknown, but it may act as an antigen receptor on the lymphocyte surface.

Antigen–Antibody Reaction Mechanism

Antigen-antibody reaction *in vitro* is known as *serological reaction* (Fig. 4-2).

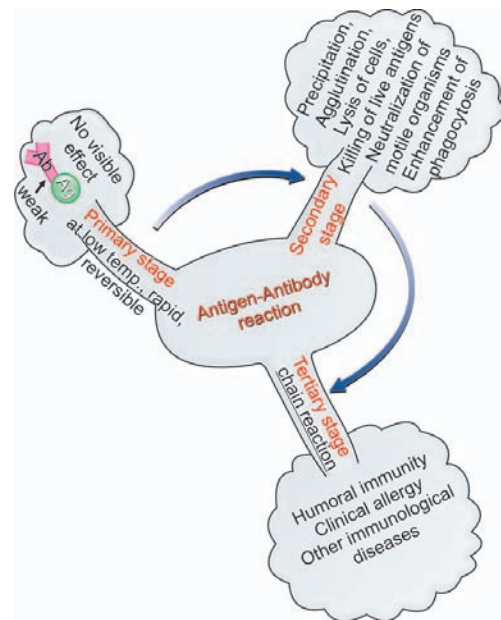


Fig. 4-2: Diagrammatic representation of antigen-antibody reaction.

- **Primary state**—In this, there is initial interaction between the two without any visible effect. This reaction is rapid and occurs even at low temperature and also is reversible because inter-molecular forces between Ag and Ab are weaker.
- **Secondary state**—it leads to precipitation, agglutination, lysis of cells, killing of live antigens, neutralization of motile organisms and enhancement of phagocytosis. An antigen can stimulate production of different types of immunoglobulins, which differ in their reaction capability and other properties.
- **Tertiary state**—some antigen-antibody reactions occurring *in vivo* initiate chain reactions that leads to neutralization or destruction of injurious antigens or tissue damage. These are tertiary reactions and include humoral immunity against infectious diseases, clinical allergy and other immunological diseases.

General Features of an Antigen-Antibody Reaction

- **Specific**—the reaction is specific; an antigen combines only with its homologous antibody and vice versa. Entire molecule reacts and not its fragments.
- **Non-degenerative**—there is no degeneration of the antigen or antibody during the reaction.
- **Combination**—the combination occurs at the surface, which is firm and reversible. Antigen and antibody can combine in varying proportions.

Precipitation Reaction

- **Formation of insoluble precipitate**—when a soluble antigen combines with antibody in the presence of electrolyte (NaCl) at a suitable temperature and pH, the antigen-antibody complex forms an insoluble precipitate which is greatly influenced by the relative proportions of antigens or antibodies.
- **Results**—if into the same amount of antiserum in different tubes, increasing quantities of antigens are added, precipitation will be found to occur most rapidly and abundantly in one of the middle tubes in which Ag and Ab are present in optimal or equivalent proportion. The precipitation will be weak in proceeding or later tubes (Fig. 4-3).

Agglutination Reaction

- **Mechanism**—when the particulate antigen is mixed with its antibody in the presence of the electrolyte at a suitable temperature and pH, the particles are clumped or agglutinated. It is more sensitive than precipitation for the detection of an antibody. It occurs optimally

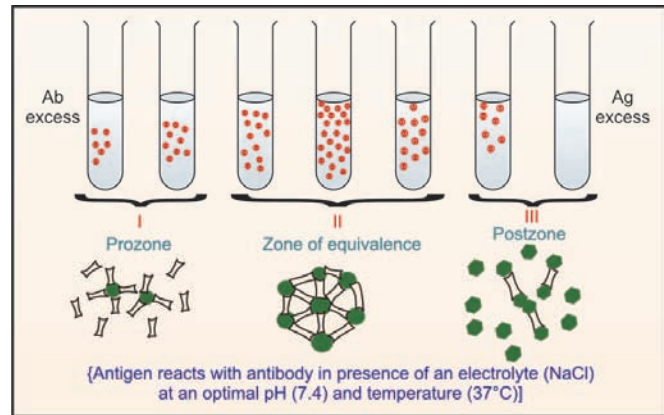


Fig. 4-3: Precipitation reaction—A diagrammatic representation.

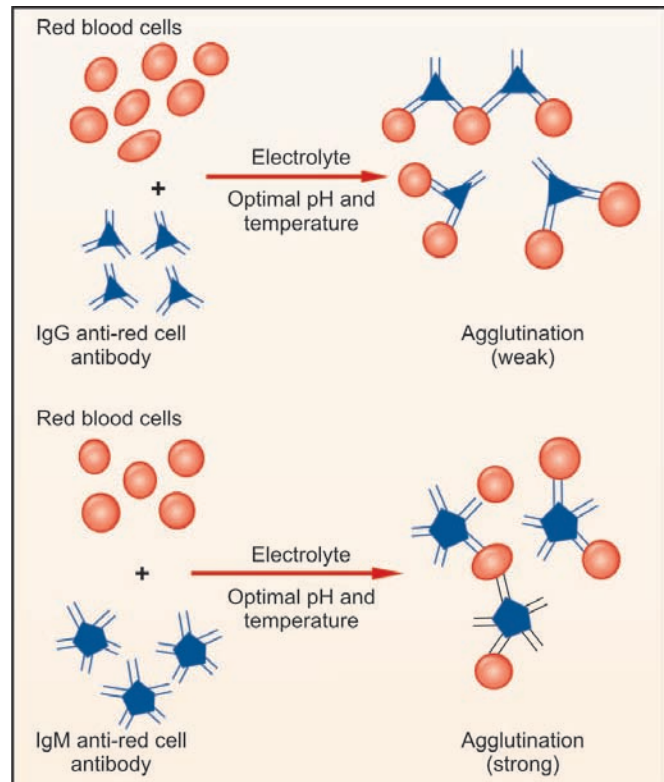


Fig. 4-4: Agglutination reaction.

when an antigen and antibody react in equivalent proportions. Better agglutination reaction takes place with IgM antibody than with IgG antibody (Fig. 4-4).

Complement Fixation Test

- **Mechanism**—complement takes part in many immunological reactions and is absorbed during the combination of antigens with their antibodies. The ability of antigen-antibody complexes to fix complement is made to use in the complement fixation test. This is a very sensitive and a versatile test.

- **Component**—it consists of five reagents which are antigen, antibody, complement, sheep erythrocytes, and amboceptor. Each of these has to be separately standardized.
- One unit or minimum hemolytic dose of complement is defined as the highest dilution of guinea pig serum that lyses one unit volume of washed sheep erythrocyte in the presence of excess of hemolysin (amboceptor) within a fixed time (usually 30 or 60 minutes) and at a fixed temperature (37°C) (Figs 4-5 and 4-6).

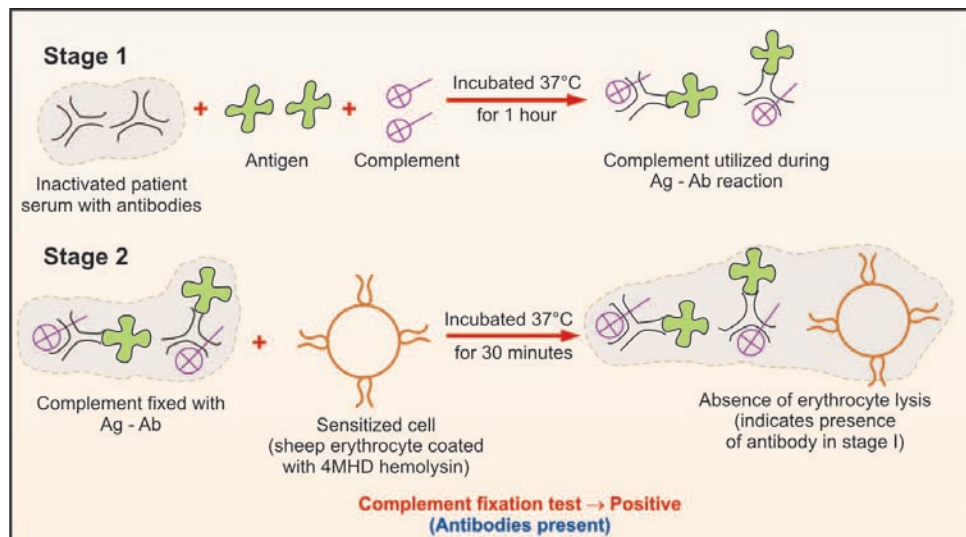
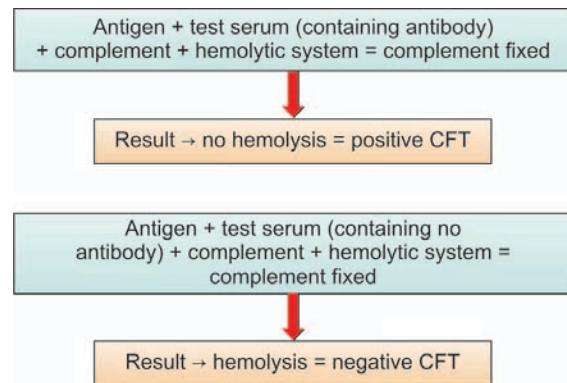


Fig. 4-5: Positive complement fixation test.

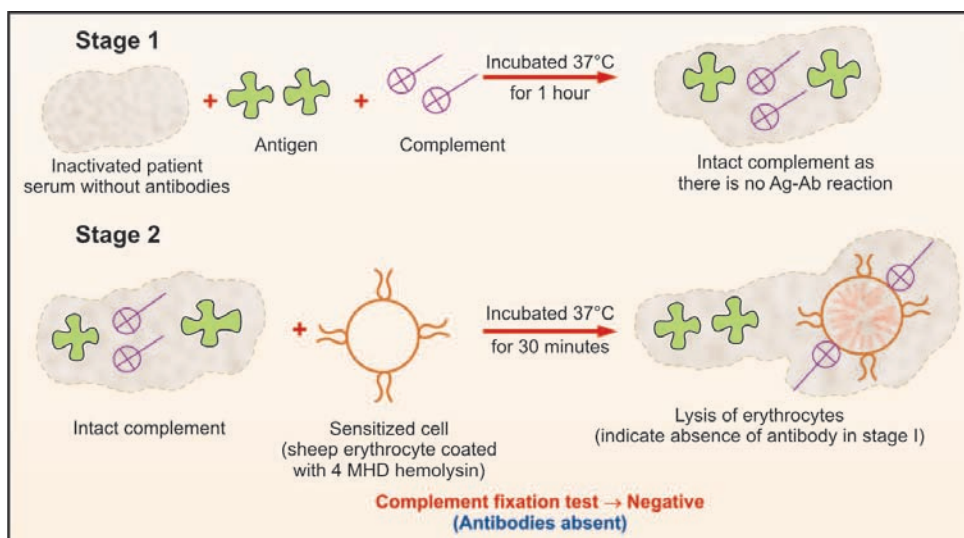


Fig. 4-6: Negative complement fixation test.

Virus Neutralization Test

Neutralization of viruses by their antibodies can be demonstrated in various systems. Neutralization of bacteriophages can be demonstrated by the plaque inhibition test. Neutralization of animal viruses can be seen in three systems: animal, eggs and tissue culture.

Immunofluorescence

- *Mechanism*—fluorescence is a property of absorbing light rays of one particular wavelength and emitting rays with different wavelengths. Fluorescence dye can be conjugated to antibodies and such labelled antibodies can be used to locate and identify antigen in tissue.

- *Use*—this test can be used for identification of bacteria, viruses or other antigens using specific antisera labelled with fluorescence dyes.
- *Dye use*—commonly used dye is fluorescein isothiocyanate and lissamine rhodamine exhibiting blue-green and orange-red fluorescence respectively.

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Neoplasm

Definition

It is an abnormal mass of tissue, growth of which exceeds and is uncoordinated with that of normal tissues and persists in the same excessive manner, after cessation of the stimuli that evokes the changes.

Oncology is the study of neoplasm.

Nomenclature

There are mainly of two types neoplasm: benign and malignant. Benign tumors are designated by attaching '*oma*' to cell of the organ. Malignant tumors arising from mesenchymal tissues are known as '*sarcomas*', like osteosarcoma. Malignant tumors of epithelial origin are called '*carcinoma*', like adenocarcinoma and squamous cell carcinoma.

Normal Cell Cycle

All renewing cells go through a series of events known as cell cycle. Successive phases of progression of cell cycle are described below (Fig. 5-1).

- *G₁ phase*—after mitosis (M phase), cells spend a variable period of resting (*G₁ phase*) where DNA synthesis is absent but the synthesis of RNA and protein continues.
- *S phase*—at the end of *G₁ phase*, unknown signals continuously institute a burst of RNA synthesis which is followed by DNA synthesis (*S phase*). Then the cells undergo replication or remain polyploid and eventually die.
- *G₂ phase*—the cells cease DNA synthesis during *G₂ phase* and DNA content is fairly constant in the growing normal cells. The proportion of cells population undergoes active proliferation in the cycle is termed as growth fraction.

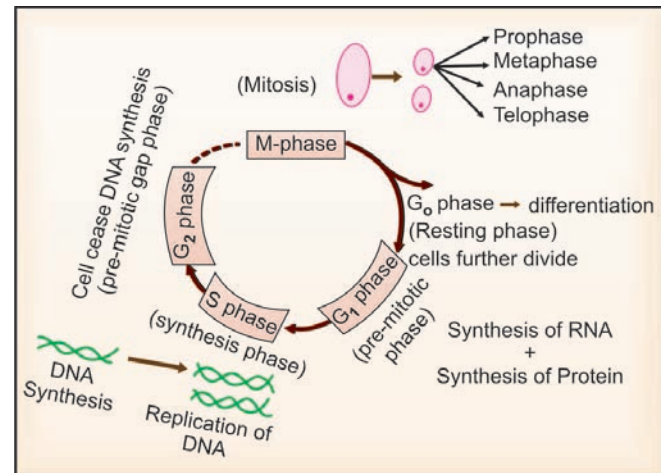


Fig. 5-1: Normal cell cycle—A diagrammatic representation.

- *G₀ phase*—daughter cells may continue to remain in cell cycle and divide further or may go out at the cell cycle in resting phase.

Predisposing Epidemiologic Factor for Development of Neoplasm

Hereditary Predisposition

The risk of developing cancer in relatives of a known cancer patient is three times higher than control study. Genetic cancers comprise not greater than 5% of all cancers, e.g. retinoblastoma, familial polyposis coli, cancer of breast etc.

Racial and Geographic Factors

Cancers are largely due to the influence of environment and geographic differences affecting whole population such as climate, water, diet, habit. For example—

- Black Africans commonly have cancers of skin, penis, cervix, and liver. Europeans and Americans commonly develop malignancies of lung, breast, and colon.

- Carcinoma of stomach is five times higher in Japanese than in Americans.
- Nasopharyngeal cancer is common in south East Asians.

Environmental and Cultural Factors

We are surrounded by an environment of carcinogens which we eat, drink, inhale and touch (Fig. 5-2). For example—

- Cigarette smoking is the etiology of cancer of oral cavity, pharynx, larynx, esophagus, lung, pancreas and urinary bladder.
- Alcohol causes cancer of esophagus and liver.
- Alcohol and tobacco together accelerate the risk of developing cancer of upper aerodigestive tract.
- Betel nut chewing causes cancer of cheek and tongue.
- Industrial and environmental materials are carcinogenic. This includes exposure to substances like arsenic, asbestos, benzene, and naphthylamine.
- Overweight individuals, deficiency of vitamin A, people consuming foods rich in animal fats and low in fiber content have more risk of developing cancers like colonic cancer.

Age

Generally, it occurs in older individuals past 5th decade of life but, there is variation in age groups. For example, acute leukemia occurs in children, neuroblastoma in infancy.

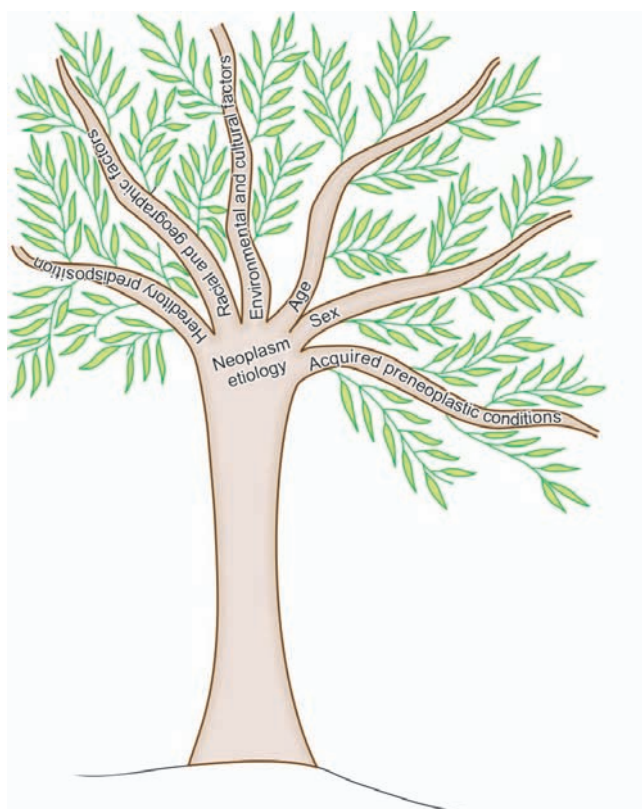


Fig. 5-2: Different types of etiological factors responsible for formation of neoplasm.

Sex

It is generally more common in men except cancer of breast, gallbladder, and thyroid.

Acquired Preneoplastic Conditions

These may be inflammatory, hyperplastic conditions or may be certain benign tumors. It includes chronic atrophic glossitis, leukoplakia of oral cavity, vulva and penis, cirrhosis of liver, chronic irritation and multiple neurofibromas.

Carcinogenesis

Carcinogenesis or oncogenesis or tumorigenesis means induction of tumors; agents which can induce tumors are called carcinogens. Carcinogens are broadly divided into four groups (Fig. 5-3):

- Chemical carcinogens
- Physical carcinogens (radiation)
- Hormonal carcinogens
- Biologic carcinogens (virus)

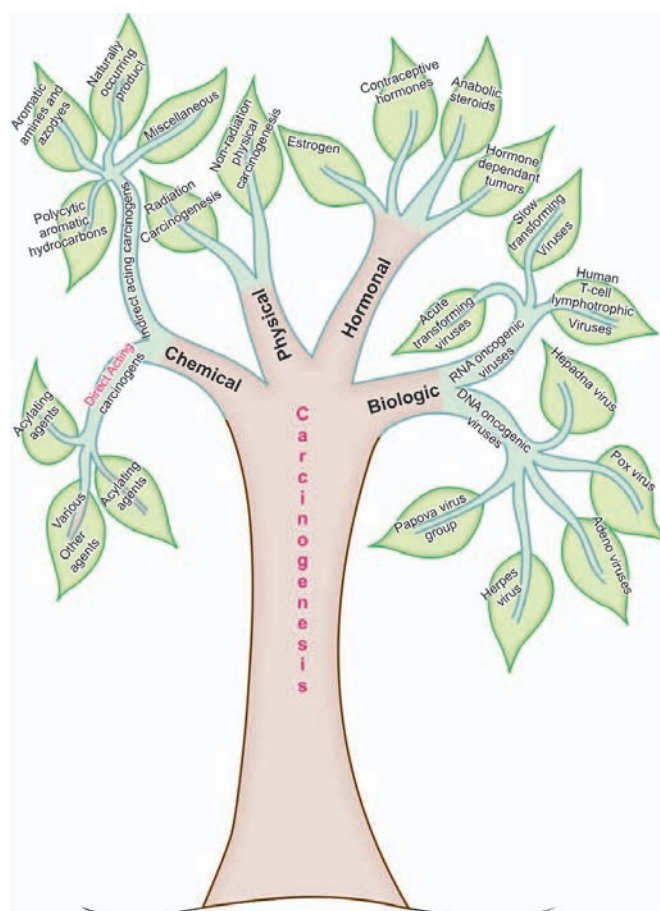


Fig. 5-3: Types of carcinogenesis.

Chemical Carcinogenesis

Chemical carcinogens are divided into two broad groups:

Initiators of Carcinogenesis

- *Direct-acting carcinogens*—these require no metabolic conversion to become carcinogens.
- *Alkylating agents*—it includes, various chemotherapeutic drugs that have successfully cured, controlled or delayed recurrence of certain types of cancers only to later evoke a second form of cancer usually leukemia. Various agents used are cyclophosphamide, chlorambucil, busulfan, melphalan, nitrosourea, β -propiolactone and epoxides. This tragic consequence is called as “*Pyrrhic victory*” which becomes less of a victory when their initial use has been converted to cause later on second form of cancer.
- *Acylating agents*—substances like acetyl imidazole.
- *Indirect-acting carcinogens or procarcinogens*—these are chemical substances requiring metabolic activation for becoming potent initial carcinogens.
- *Polycyclic aromatic hydrocarbons* (in tobacco, smoke, animal foods, industrial oil, and atmospheric pollutants). Important chemical compounds included are benzanthracene, benzapyrene, methylcholanthrene. They may cause lung cancer, skin cancer, cancer of oral cavity, and sarcoma.
- *Aromatic amines and azo dyes*—These are β -naphthylamine, benzidine, azo dyes used for coloring foods, and acetyl aminofluorene. They may cause bladder cancer and hepatocellular carcinoma.
- *Naturally occurring product*—Aflatoxin, actinomycin-D, mitomycin-C, safrole, and betel nut. It can cause hepatocellular carcinoma.
- *Miscellaneous*—nitroso compounds, vinyl chloride monomer, asbestos, arsenical compounds, metals like nickel, lead, chromium, and insecticides, fungicides can cause gastric carcinoma, hemangiosarcoma of liver, bronchogenic carcinoma, epidermal hyperplasia, basal cell carcinoma, and lung cancer.

Promoters of Carcinogenesis

Certain chemical substances lacking the intrinsic carcinogenic potential but helping the initiated cell to proliferate further are called promoter of carcinogenesis. E.g. phorbol, phenols, drugs like phenobarbital, and artificial sweetener like saccharine.

Mechanism of Action

- *Binding to DNA and RNA*—the great majority of chemical carcinogens are mutagens. They bind directly to DNA and RNA or cytoplasmic proteins to specific sites within

molecule inducing miscoding error during transcription and replication.

- *Factors affecting a carcinogenicity*
 - *Dose dependent*—the carcinogenicity of chemical agents is dose dependent and multiple traditional doses have same oncogenicity as a single comparative dose.
 - *Administration of promoters*—the carcinogenicity of chemical agents can be significantly enhanced by the subsequent administration of promoters. To be effective, the promoter must follow the initiator.

Stage of Chemical Carcinogen

The phenomenon of cellular transformation by chemical carcinogenesis is a progressive process involving two different stages. These are initiation and promotion.

Initiation

In this initiator carcinogen interact with DNA of target cell to induce mutation that is more or less irreversible to transform it into initiated cell.

- *Metabolic activation*—only indirect acting carcinogen or procarcinogens require metabolic activation chiefly by mixed oxidases of cytochrome P-450 system located in microsomal compounds of the endoplasmic reticulum or in the nucleus.
- *Reactive electrophiles*—they are electron deficient protons, which bind to electron rich portions of other molecules of cell such as DNA, RNA or other protein.
- *Target molecules*—the primary target is DNA, producing mutagenesis.
- *Initiated cell*—the unrepaired damage produced in the DNA of the cell becomes permanent, only if the altered cell undergoes at least one cycle of proliferation.

Promotion

It does not damage the DNA but enhances the effect of direct-acting carcinogen or procarcinogens. The ultimate effect is further clonal proliferation of the initiated cell. Two or more initiators may be chemical, oncogenic virus or radiant energy may act in concert to induce malignant transformation referred to as cocarcinogens.

Physical Carcinogenesis

It is divided into two groups.

Radiation Carcinogenesis

- *Forms of radiation*—radiation whenever in the form of UV light from sunlight, UV lamp, welder's arc, or ionizing radiation like X-ray, α , β and γ ray, radioactive isotopes, protons and neutrons are established carcinogens.
- *Example of radiation induced cancers*—most frequent radiation induced cancers are leukemia, cancer of

thyroid, skin, breast, lung, and salivary gland. Therapeutic irradiation can also induce carcinogenesis.

- *Facts about radiation causing cancer*—radiant energy have potential of producing mutation and even killing cells. It can affect carcinogenesis by the following facts:
 - Few tumors appear only after long latent period during which successive generation of clones are developed.
 - The radiation initiation is generally irreversible, but at a low dosage level is amenable to repair.
 - The effect of radiation depends upon a number of factors such as type of radiation, dose, length of interval between the doses, capability of cells to repair in intervals and various host factors such as age, individual susceptibility, immune competence, hormonal influence and type of cell irradiated.
- *Mechanism*—it induces cancer by following mechanism;
 - Radiation may directly alter the cellular DNA and it may dislodge ions from water and other molecules of cell and result in the formation of highly reactive free radicals that may bring about the damage.
 - Radiation mutation may render cell vulnerable to other carcinogenic influence, i.e. acting as co-carcinogen.
 - Inhibition of cell division and inactivation of enzymes.
 - Radiation might cause cell killing; permitting survivors to proliferate and thereby, become vulnerable to oncogenic influence.

Non-radiation Physical Carcinogenesis

Mechanical injury to tissues such as from stones in the gall bladder, stones in the urinary tract, and healed scars following burns or trauma has been suggested as causes of increased risk of carcinoma.

Implants of inert materials such as plastic, glass, etc. in prosthesis and foreign bodies like metal foils observe to cause tumor development in experimental animals.

Hormonal Carcinogenesis

Carcinoma is most likely to develop in organs and tissues which undergo proliferation under influence of excessive hormonal stimulation. Hormone sensitive tissues developing tumors are breast, endometrium, myometrium, vagina, thyroid, liver, prostate, and testis.

- *Estrogen*—in experimental animals, estrogen can cause induction of breast cancer in mice. Other cancers which can be induced in mice by estrogens are squamous cell carcinoma of cervix, connective tissue tumor of myometrium, tumor of kidney in hamsters, and benign and malignant tumors of liver in rats. In case of human women receiving estrogen therapy and women with estrogen secreting granulosa cell, tumor of the ovary have

increased risk of developing endometrial carcinoma. Adenocarcinoma of the vagina is seen with increased frequency in adolescent daughter of mother who had received estrogen therapy during pregnancy.

- *Contraceptive hormones*—there is increased risk of developing breast cancer, benign tumors of the liver and few patients have developed hepatocellular carcinoma.
- *Anabolic steroids*—consumption of anabolic steroids by athletes to increase the muscle mass also increases the risk of developing benign and malignant tumor of the liver.
- *Hormone dependent tumors*—it has been shown in experimental animals that induction of hyperfunction of adeno-hypophysis is associated with increased risk of developing neoplasia of the target organs following preceding functional hyperplasia.

Biologic Carcinogenesis

The epidemiological studies on different types of cancer indicate the involvement of transmissible biologic agents in their development, chiefly viruses. It has been estimated that about 20% of all cancers worldwide are virus associated cancers. Therefore biological carcinogenesis is largely viral oncogenesis. A large number of viruses have been proved to be oncogenic in wide variety of animals and in certain types of cancers in humans.

The association of oncogenic virus with neoplasia was observed by an Italian physician Sanarelli in 1889 who noted association between myxomatosis of rabbit with poxvirus. Oncogenic viruses fall into 2 broad groups, i.e. those containing ribonucleic acid are termed as RNA oncogenic viruses and those containing deoxyribonucleic acid are termed as DNA oncogenic viruses.

RNA Oncogenic Viruses

These are retroviruses, i.e. they contain the enzyme reverse transcriptase, which is required for reverse transcription of viral RNA to synthesize viral DNA strands. Based on their activity to transplant target cells into neoplastic cells, they all are divided into three subgroups:

- *Acute transforming viruses*—it includes Rous sarcoma virus in chickens, leukemia-sarcoma viruses of avian, feline, bovine and primate.
- *Slow transforming tumor viruses*—mouse mammary tumor virus (MMTV) that causes breast cancer in daughter mice.
- *Human T-cell Lymphotropic viruses (HTLV)*—it can cause adult T-cell leukemia-lymphoma syndrome and AIDS.

DNA Oncogenic Viruses

They are divided into five groups

- *Papovavirus group*—human papilloma virus, polyoma virus, SV-40 (simian vacuolating) virus.

- *Herpes virus*—Epstein Barr virus, Human herpesvirus, cytomegalovirus, Lucke's frog virus, Marek's disease virus.
- *Adenoviruses*—it can cause upper respiratory infections and pharyngitis. In man, they are not known to be involved in tumors but in hamsters they may induce sarcomas.
- *Poxvirus*—in rabbits, it can cause myxomatosis and in humans, it can cause molluscum contagiosum and may induce squamous cell papilloma.
- *Hepadnaviruses*—hepatitis B virus is a member of this family and it can cause acute hepatitis and is responsible for carrier state which can result chronic hepatitis. In some cases, to progressing to hepatic cirrhosis and into hepatocellular carcinoma.

Mechanism of Biological Carcinogenesis

RNA viral oncogenesis:

Reverse transcriptase acts as a template to synthesize a single strand of matching viral DNA.

- Single strand of viral DNA is then copied by DNA dependent DNA synthetase to form another strand of complementary DNA resulting in double stranded viral DNA or provirus.
- The provirus is then integrated into the DNA of the host cell genome and may transform the cell into a neoplastic cell.
- Virus replication begins after integration of provirus into host cell genome. Integration results in transcription of proviral genes or progenes into messenger RNA which then forms components of the virus particle, i.e. virion core proteins from gag gene, enveloped glycoprotein from env gene and reverse transcriptase from pol gene.
- The three components of virus particles are then assembled at the plasma membrane of host cells and virus particles released by budding off from plasma membrane, thus completing the process of replication.

DNA viral oncogenesis:

- *Replication*—The virus may replicate in the host cell with consequent lysis of infected cell and release of virions.
- *Integration*—The viral DNA may integrate into the host cell DNA. This results in neoplastic transformation of the host cell.

Oxidative Mechanism of Carcinogenesis

Active oxygen species and other free radicals have long known to be mutagenic. Further, these agents have emerged as mediators of the other phenotypic and genotypic changes that lead to form mutation to neoplasia.

Free radicals production is ubiquitous in all respiring organism and is enhanced by many disease states, by

carcinogen exposure and under conditions of stress. Free radicals may therefore contribute widely to cancer development in humans.

Free radicals scavenging vitamins C and E have been shown to protect against cancer development in animal models.

Biology of Tumor Growth

The life cycle of malignant tumors can be divided into four phases.

Induction of Malignant Changes in the Target Cell (Transformation) (Fig. 5-4)

- *Effect on genes*—large number of carcinogen agents induce neoplastic transformation of cells *in vivo* and in experimental animals. All etiologic factors ultimately affect the function of two sets of genes, one is proto-oncogenes or oncogenes and another one is *anti-oncogenes* or cancer suppressor genes.
- *Binding with DNA*—the majority of carcinogens are mutagens which bind the DNA directly or indirectly by undergoing enzymatic activation, inducing miscoding errors during transcription and replication.
- *Production of growth factors*—oncogenes may code for growth promoting factors and as a result, the tumor cells produce large amount of growth factors to which, only they can respond.

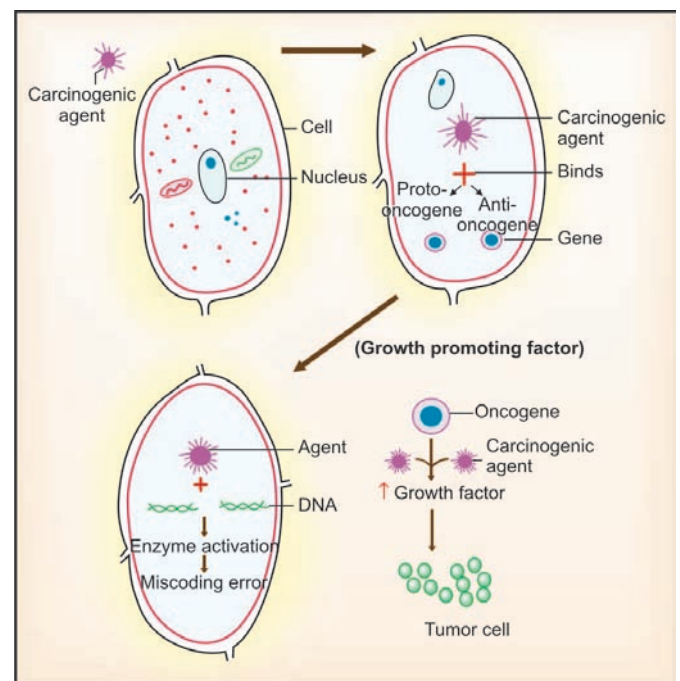


Fig. 5-4: Biology of tumor growth (induction phase).

- **Encoding**—oncogenes may encode a defective receptor that send stimulating signals to the cells, even in the absence of growth factors.
- **Multiple mutations**—cancer is a genetic disease that results when multiple mutations accumulate in the DNA of a cell and specific chromosomal abnormalities predispose to cancer.

Growth of Transformed Cells (Kinetics of Tumor Cell Growth) (Fig. 5-5)

- **Doubling**—the monoclonal cancer cell (10 μm in diameter) has to undergo about 30 population doublings to produce 10^9 cells weighing approximately 1 gm, which is the smallest clinical detectable mass. To produce a tumor of 10^{12} cells, weighing 1 kg approximately, which is usually the maximum size compatible with life, the tumor cells have to undergo 10 further population doublings. So by the time the tumor is clinically detectable, it has already complete a major portion of its life cycle.
- **Factors affecting growth of cells**—in tumor cells, there is an imbalance between cell production and cell loss, therefore, the tumor grows progressively. The rate of tumor growth depends upon the growth fraction and the degree of imbalance between cell production and cell loss.

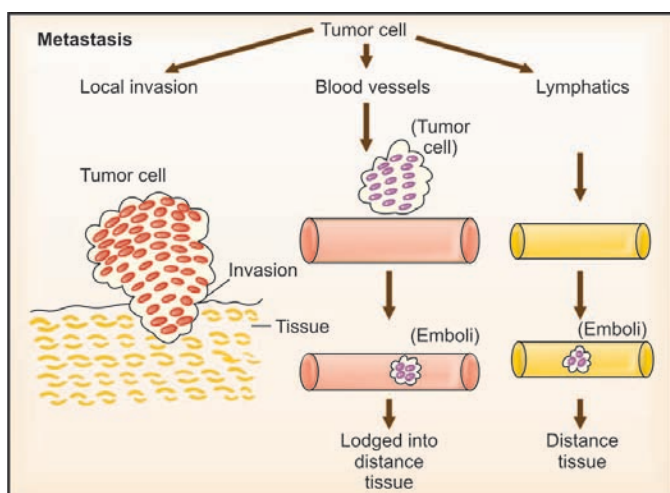


Fig. 5-5: Biology of tumor growth (kinetic of tumor cell growth).

Mechanism of Local Invasion and Distant Metastases

- **Routes of metastasis**—there are three routes through which metastases of tumor cells occur, i.e. local invasion, via blood vessels and via lymphatics.
- **Local invasion**—the local invasion takes the path of least resistance and the tumor cells invade the surrounding tissue spaces.

- **Lymphatics spread**—in case of oral malignancies, distant metastasis is mainly via lymphatics, either by lymphatic permeation or by lymphatic embolism.
- **Blood vessels**—rarely, it spreads through blood vessels and if this occurs, the tumor cells invade the lumen of blood vessels, the tumor emboli form, which are fragmented and the tumor cells are lodged into distant tissues.

Theories of Carcinogenesis

The Epigenetic Theory

According to this theory, the carcinogenic agents act on the activators or suppressors of genes and not on the genes themselves and result in the abnormal expression of genes.

Genetic Theory

- **Concept**—this is the most popular theory which suggests that cells become neoplastic because of alteration in the DNA. It is suggested that, the secret of cancer lies within the normal cells themselves in the form of proto-oncogenes (C-oncs). The mutated cells transmit their characters to the next progeny of cells. Inappropriate over expression of the gene or point mutation cause the cell to produce stimulating growth factors or in some way damages normal regulatory control. The qualitative and quantitative changes in the expression of genome may be brought about by carcinogenic influence, i.e. chemical, viruses, radiation or spontaneous random mutations.
- **Oncogenes**—Oncogenes are the transforming genes present in many tumor cells. Closely related genes are detected on normal animal and human cells and are called as 'proto-oncogenes' or 'cellular oncogenes', abbreviated as 'c-oncs'.
- **'Cellular oncogenes'** of the host cells can transcribe its copies in the viral genome of acute transforming oncogenic retroviruses called as viral oncogenes or '*v-oncs*'. An alternate mechanism is by anti-oncogenes in which, there is inactivation or deletion of genes that normally perform the function of suppressing cell proliferation, thus allowing them to proliferate.
- **Feedback deletion**—according to genetic regulatory mechanism theory, primary change in the cell consists of a modification of repressor molecule which controls the functions of the gene. The repressor molecules are either RNA or protein. The modification of repressor molecules removes their orderly inhibitory control, which is responsible for normal morphogenesis and differentiation, and unearths the cell genetic potentiality for unrestricted growth. This concept of loss of growth control is described as '*feedback deletion*'.

Virus Theory

At some stage, it participates in the development of cancer. The concept of mode of action of virus has taken many forms-

- *Act as parasite*—virus is present as a parasite in all tumor cells and it is transmitted from cell-to-cell and stimulates extreme hyperplasia without affecting the genome cell.
- *Act as biologic carcinogen*—it acts as a biologic carcinogen on some cellular constituents to release or activate neoplastic potentialities normally present in cells.
- *Auto synthesis*—carcinogens of all kinds ultimately act by creating some new auto-synthesizing cytoplasmic constituents, probably an auto-catalytic protein, which can excite the cell to unlimited growth.

Immune Surveillance Theory

It suggests that an immune-competent host mounts an attack on developing tumor cells so as to destroy them while an immune-incompetent host fails to do so. According to original immunological theory, normal cells contain specific 'self-marker' (identity proteins) which is recognized by the normal growth regulating mechanism.

These proteins serve as receptor for chemical carcinogens (hapten) and the resulting complex is self-replicating. The complex (complex antigen) triggers off an immune response and the antibody (free or cell bound) combines with the self-marker carcinogen complex and eliminates it.

The new race of cells produced is deleted with self-markers and goes unrecognized by growth regulatory mechanism. The high incidence of cancer in AIDS patients is in support of this theory.

Monoclonal Hypothesis

Currently, there is strong evidence on studies of human and experimental animals that most of the cancers arise from single clone of transformed cell.

The best documentation of monoclonal origin of cancer cells come from the study of G₆PD in women who are heterozygous for its two isoenzymes A and B. It is observed that all tumor cells in benign uterine tumor (leiomyoma) contain either A or B genotype of G₆PD—i.e. the tumor cells is derived from a single progenitor cell.

Multistep Theory

According to this theory, carcinogenesis is a multistep process which is substantiated *in vitro* by changes in experimental animals as well as *in vivo* by changes in human cancers.

In chemical carcinogenesis, there are two essential features, i.e. initiation and promotion. Many tumors arise from combination of activation or growth promoting

oncogenes and inactivation of growth suppressing anti-oncogenes. In some cancers, there is initial dysplastic change that may progress into carcinoma *in situ* and then into invasive carcinoma.

Metastasis

'Metastasis is defined as spread of tumor by invasion in such a way that discontinuous secondary tumor mass/masses are formed at the site of lodgment'. This metastasis is the transfer of the disease from one organ or part to another not directly connected with it.

If malignant cells did not metastasize, the surgical removal of primary neoplasm would completely cure the patient. Metastasis is fundamentally an embolic process. The invasiveness of malignant cells involves motility, which requires change in shape and adhesiveness and ability to degrade the matrix in order to penetrate it.

Thus, a definition of the behavior of the metastatic tumor cells is the tendency to cross the tissue compartment / boundary and intermix with other cell types. The metastatic process can be divided into several sequential steps although these steps are interconnected.

Factors which control metastasis are proteolysis, cell adhesion, tumor angiogenesis, cell mediated immunity and genetic factor.

Steps of Metastasis

- *Breaking of cells*—the breaking of loose neoplastic cells from the parent tumor.
- *Invasion*—invasion of the matrix (sarcoma), penetration of the basement membrane and invasion of connective tissue (carcinoma).
- *Entering the blood vessels*—it then entering the wall of blood and lymphatic vessels.
- *Survival*—survival of malignant cells in the bloodstream. Survival in the compatible tissue environment and induction of growth factor to stimulate new vessel formation to obtain nutrition.
- *Emergence of cells*—emergence of the malignant cells from the blood vessels in the form of the emboli and lodgment in other tissues.
- *Multiplication*—multiplication of neoplastic cells and growth to form secondary neoplasm at the new site.
- *Control mechanism*—each of these steps is probably controlled by different molecular mechanism and this may explain the differences in the behavior with reference to tumor metastasis. Neoplastic cells within a single tumor might differ in their ability to metastasize. A sub-population of cells preexists within the heterogeneous primary tumor. The relative size of this sub-population in the primary tumor may vary with time between the neoplasms.

Routes of Metastasis (Fig. 5-6)

- *Lymphatics*—particularly for carcinoma and lymphosarcoma. For example, mouth-to-neck nodes and breast-to-axillary nodes.
- *Bloodstream*—particularly veins from gut via portal circulation to liver, from systemic sites through right heart to lung, from left heart to any systemic sites.
- *Cavities*—along epithelium lined cavities, for example: respiratory tract, gut, urinogenital tract etc.
- *Others*—transcelomic spread, cerebrospinal fluid, tissue planes and through nerve sheath.

Pattern of Metastatic Spread

- *Mechanistic theory*—the capillary bed of the first organ which encounter viable neoplastic cells is the preferred site of metastasis.
- *Seed and soil hypothesis*—it suggests that availability of fertile environment (the soil) in which compatible tumor cells (the seed) can grow is important. Ewing suggested that varying pattern of metastasis is due to fact that different tumor cells thrive in certain biological sites (soils) but not in the other sites.
- *Cell interaction*—interaction between cell surface protein of malignant cells and organ specific protein, e.g. fibronectin receptor.

Grading and Staging of Tumors

Grading

The grading features are those indicative of proliferation and differentiation. It is defined as, 'macroscopic and microscopic degree of differentiation of tumor'. Grading depends mainly on two histologic features, the degree of anaplasia and the rate of growth. Different types of grading systems are as follows:

Broder's Classification System (Fig. 5-7)

- *Grade I* (well differentiated, i.e. less than 25% anaplastic cells)—it is characterized by the presence of relatively mature cell with few nuclear aberration and with the presence of keratin pearls and individual cell keratinization.
- *Grade II*—(Moderately differentiated, i.e. 25 to 50% anaplastic cells)—it is characterized by the presence of tumor cell exhibiting a wide range of differentiation, keratinization is occasionally present, and nuclear aberrations are moderately abundant. Usually, the invasion is poorly delineated from the stroma.
- *Grade III*—(Moderately differentiated, i.e. 50 to 75% anaplastic cells)—it is characterized by disorderly and poorly differentiated cells with no tendency towards keratinization, nuclear aberrations are abundant.

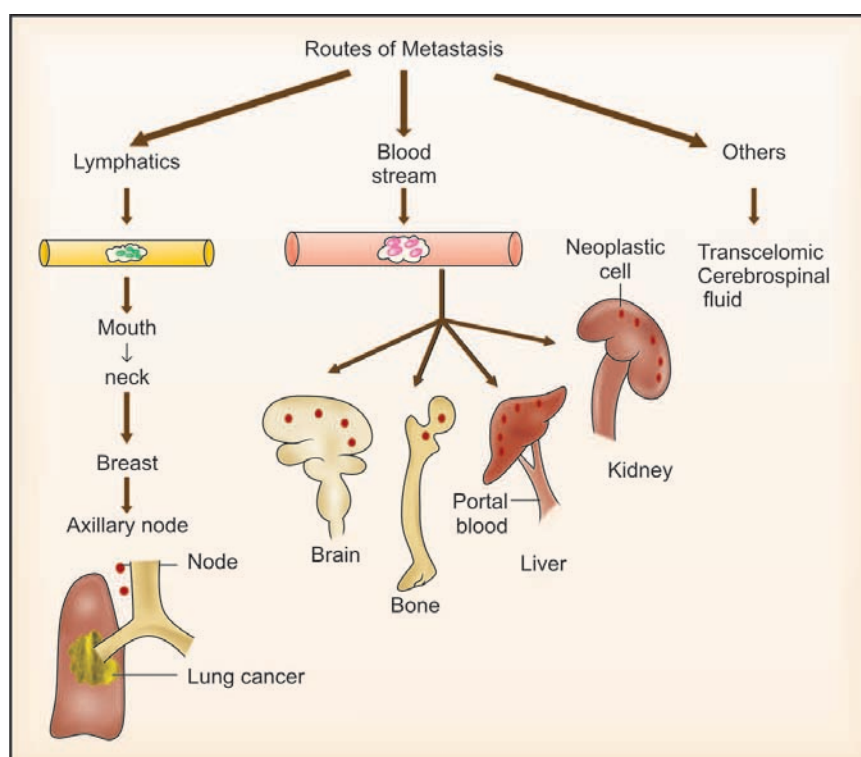


Fig. 5-6: Different routes of metastasis in the human body.

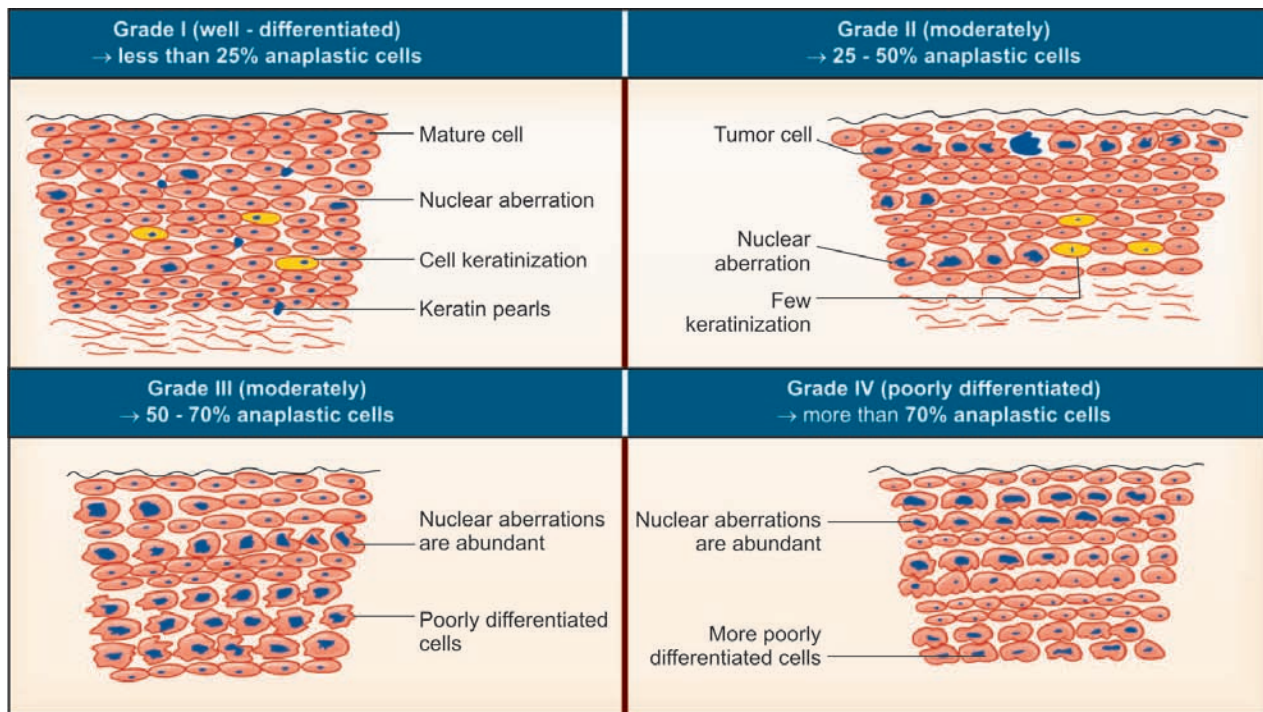


Fig. 5-7: Broader's classification (Grading System)—A diagrammatic representation

- **Grade IV**—(Poorly differentiated, i.e. more than 75% anaplastic cells)—in it, cells are so poorly differentiated that they cannot be identified as epithelial origin on the basis of histology alone, nuclear aberrations are abundant and no keratinization is found.

CIN Grading

Alternative classification for grading of dysplasia and carcinoma *in situ* together is cervical intraepithelial neoplasia (CIN). According to this grading, the criteria are as follows:

- **CIN I**—it represents less than 1/3rd involvement of the thickness of the epithelium.
- **CIN II**—in it, there is 1/3rd to 2/3rd involvement.
- **CIN III**—it is full thickness or equivalent to carcinoma *in situ*.

Depending on thickness of squamous epithelium involved by atypical cells

According to this, grading of dysplasia is as follows:

- Mild
- Moderate
- Severe

Staging

It is the extent of spread of tumor within patients.

Assessment

It is assessed by following ways:

- **Clinical examination**—staging can be assessed clinically by the size and extent of primary lesion.
- **Investigation**—investigation like radiology, sonography can lead to staging of disease.
- **Pathological examination**—this is very important part while staging of neoplastic process is done. Mainly biopsy, cytology are used.
- **Infiltration**—degree of infiltration of primary lesion should be carried out.
- **Metastasis**—presence or absence of metastasis to regional lymph nodes. Presence and absence of distant metastasis. Involvement of contralateral or ipsilateral node. Whether node are fixed or not.

Objectives

- **Treatment planning**—it aids clinician in the planning of treatment.
- **Prognosis**—to give some indication of prognosis.
- **Evaluation of treatment**—to assist in evaluation of the result of treatment.
- **Exchange of information**—to facilitate the exchange of information between treatment centers.
- **Investigation**—to contribute to the continuing investigations of human cancer.

TNM Staging

It is universally accepted system which is developed by UICC (Union Internationale Contre of Cancer).

- *Primary tumor (T)*: local extent is major factor contributing to prognosis.
 - T_x: primary tumor cannot be assessed.
 - T₀: no evidence of primary tumor.
 - T_{is}: carcinoma *in situ*.
 - T₁: tumor 2 cm or less in diameter.
 - T₂: tumor 2-4 cm in diameter.
 - T₃: tumor more than 4 cm in greatest diameter.
 - T₄: tumor of any size in which tumor invades adjacent structure (e.g.: cortical bone, inferior alveolar nerve, floor of mouth, skin of face etc).
- *Regional lymph nodes (N)*
 - N_x: regional lymph node cannot be assessed.
 - N₀: no regional lymph node metastasis.
 - N₁: metastasis in single ipsilateral lymph node less than 3 cm in diameter.
 - N_{1a}: nodes considered not contain to tumor growth.
 - N_{1b}: nodes considered to contain growth
 - N₂: single lymph node, no more than 6 cm in greatest dimension, of bilateral / contralateral lymph node, no more than 6 cm.
 - N_{2a}: Single ipsilateral lymph node more than 3 but less than 6 cm.
 - N_{2b}: multiple ipsilateral lymph nodes less than 6 cm.
 - N_{2c}: Bilateral or contralateral lymph node less than 6 cm in greatest dimension.
 - N₃: metastasis in lymph node more than 6 cm and it is fixed.
 - N_{3a}—ipsilateral nodes at least one greater than 6 cm.
 - N_{3b}—bilateral nodes greater than 6 cm.
 - N_{3c}—contralateral nodes at least one greater than 6 cm.
- *Distant metastasis (M)*
 - M_x: Distant metastasis cannot be assessed.
 - M₀: No distant metastasis.
 - M₁: Distant metastasis.
 - Category M₁ may be further specified according to the notation.
 - Pulmonary—PUL
 - Osseous—OSS
 - Hepatic—HEP
 - Brain—BRA
 - Lymph nodes—LYM
 - Bone marrow—MAR
 - Pleura—PLE
 - Peritoneum—PER
 - Skin—SKI
 - Other—OTH

AJC (American Joint Committee)

It divides all cancers into stage 0 to 4, and takes into account all three previous TNM systems.

- Stage 0—T₀ N₀ M₀
- Stage 1—T₁ N₀ M₀
- Stage 2—T₂ N₀ M₀
- Stage 3—T₃ N₀ M₀, T₁ N₁ M₀, T₂ N₁ M₀, and T₃ N₁ M₀
- Stage 4A—T₄ N₀ M₀, T₄ N₁ M₀, any T N₂ M₀
- Stage 4 B—Any T N₃ M₀
- Stage 4 C—Any T, Any N, M₁

Dukes ABC Staging

It is used in cancers of bowel

- *Stage A*—when tumor is confined to submucosa and muscle and cure rate is 100%.
- *Stage B*—tumor penetrates the entire thickness of bowel wall into pericolic or perirectal tissues and cure rate is 70%.
- *Stage C*—it is characterized by lymph node metastasis and reduces the cure rate to 30%.

STNMP Staging System

- *S—site of primary tumor*
 - S₁—lip and skin.
 - S₂—lip mucosa.
 - S₃—tongue.
 - S₄—cheek.
 - S₅—palate.
 - S₆—floor of mouth.
 - S₇—alveolar process.
 - S₈—antrum.
 - S₉—central carcinoma of bone.
- *Size of tumor—it is denoted by T*
 - T₁—less than 2 cm in diameter.
 - T₂—between 2 cm and 4 cm in diameter.
 - T₃—between 4 cm and 6 cm in diameter and extending beyond the primary region and extending through adjacent periosteum.
 - T₄—greater than 6 cm in diameter and extending to involve adjacent structures.
- *Regional nodes were grouped as*
 - N₀—no palpable nodes.
 - N₁—equifocal node enlargement.
 - N₂—clinically palpable homolateral regional nodes, not fixed.
 - N₃—same as N₂ but fixed.
 - N₄—clinically palpable contralateral or bilateral nodes, not fixed.
 - N₅—same as N₄ but fixed.
- *Metastasis*
 - M₀—no distant metastasis.

Differences between benign and malignant tumors

Features	Benign tumors	Malignant tumors
Macroscopic Features		
Boundaries	Encapsulated or well circumscribed	Poorly circumscribed and irregular
Surrounding tissues	Often compressed	Usually invaded
Size	Usually small	Often large
Growth	Slow and expanding	Rapid and infiltrating
Capsule	Present	Absent
Degeneration	Rare	Common
Recurrence	Not common	Common
Fixity	Absent	Present
Secondary changes	Occur less often	Occur more often
End of growth	May come to standstill	Growth rarely ceases
Microscopic features		
Pattern	Usually resemble tissue of origin	Have poor resemblance to tissue of origin
Basal polarity	Retained	Lost
Pleomorphism	Usually not present	Often present
Nucleo-cytoplasmic ratio	Normal	Increased
Anisonucleosis	Absent	Present
Hyperchromatism	Absent	Often present
Mitosis	May be present but always with typical mitosis	Mitotic figures are increased and are generally atypical and abnormal
Tumor giant cells	May be present but without nuclear atypia	Present with nuclear atypia
Cytoplasm	May show normal constituents	Normal cytoplasmic elements are decreased or lost
Function	Well maintained	May be retained, lost or become abnormal
Growth rate	Usually slow	Usually rapid
Local invasion	Often compresses the surrounding tissue without invading or infiltrating	Usually infiltrates and invades the adjacent tissue
Metastasis	Absent	Present frequently

- M₁—clinical evidence of distant metastasis without definite histological or radiographic conformation.
- M₂—proven evidence of metastasis beyond regional nodes.
- *Pathology of lesion*
 - P₀—hyperkeratotic lesion showing atypia.
 - P₁—carcinoma in situ.
 - P₂—basal cell carcinoma.
 - P₃—verrucous carcinoma.
 - Well differentiated sq. cell carcinoma.

- Moderately differentiated sq. cell carcinoma.
- Poorly differentiated sq. cell carcinoma.

Suggested Reading

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6

Infection Control in Dental Office

Introduction

In dental practice, the risk of infection is a high priority issue, considering the nature of oral environment which is rich in diverse aerobic and anaerobic bacterial flora and more hazardous viral pathogens. Dental personnel and patients are at increased risk of acquiring many diseases which can get transferred from patient to doctor and vice versa. With changing times, risk associated with viral pathogens such as herpes, hepatitis and HIV are increased. The common goal of infection control is to eliminate or reduce the number of microbes shared between people.

Many objects in the dental office are potential source of infection. These are called as '*vectors*'. For examples; saliva, blood, nasal secretion, dust, hands, hair, clothing, films, X-ray machine, dental instrument and dental chairs. Cross infection occurs when these vectors transmit pathogenic organisms from one patient to another or to dental personnel.

All patients should be treated using universal precaution. There should be no exception and no extra precaution for specific patients. By practicing infection control, patients as well as operator can be protected.

High standard of dental infection control and occupational safety must be followed by dental personnel; for the safety of patient and dental health care workers. The word infection control does not mean total prevention of iatrogenic, nosocomial or occupational exposure, but it means to reduce the risk of disease transmission.

General Consideration

Every person working in hospital or general practitioner has a legal duty to ensure that all necessary steps are taken to prevent cross infection protect themselves, their colleagues and the patients.

Effective infection control measures are required in dental radiography even though most investigations are non-invasive or non-exposure prone and they do not involve breaches of the mucosa or skin.

Every patient in the department should have current medical history. Information gained by the history will alert the dental team about high risk situation.

In dentistry, infection can occur due to any of the following:

- *Salivary contamination*—the main risk from cross infection is due to salivary contamination from one patient to another or from working areas and equipments.
- *Contaminated object*—indirect contact with contaminated objects or instrument can occur.
- *Airborne infection*—direct contact with airborne contaminant present in spatter or aerosols or oral and respiratory fluids.

Identification of the risk and the presence of these infections is achieved by recording and keeping updated medical history. But, this cannot be relied on as:

- Patient may be unaware that they are infected. 50% of hepatitis B cases are asymptomatic and up to 10% become carrier.
- Patients may not have understood the diagnosis given to them by their medical practitioner.
- Patient may be incubating the infection.
- Information may be deliberately withheld.

It is therefore safer for HCW (health care worker) to accept that all patients can be infectious and age or class of the patient is no excuse.

Routes of Infection Transfer (Fig. 6-1)

- *Percutaneous*—this is high risk route. In this, transfer of microbes from saliva and blood can occur through needles and sharp instruments.

- **Contact**—this is also high risk transmission. In this, touching or exposing non-intact skin to infective oral lesion, infected tissue surface or infected fluids, splash and spatter of infected fluids.
- **Inhalation**—this is moderate risk transmission. In this, inhalation of aerosol or droplet containing pathogen can occur. Pathogens suspended in air from handpiece and droplet nuclei from coughing.
- **Through fomites**—it is low risk. It is very rare to touch contaminated inanimate surface in dental room or operation.

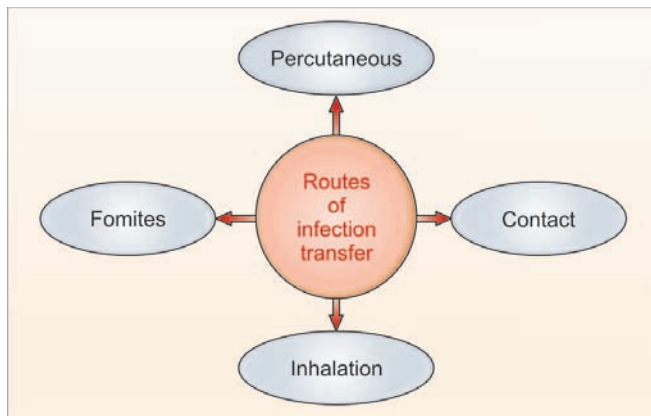


Fig. 6-1: Different routes of infection transfer

Infection of Main Concern

- **Infective hepatitis**—it includes mainly hepatitis B or hepatitis C. Risk group include IV drug user, heterosexual with multiple partners, homosexual, mentally subnormal patients and hemodialysis patient. 2% of the populations are known carriers of HBV which can be transmitted by saliva.
- **HIV and AIDS**—It is caused by HIV virus. Risk group include homosexuals, heterosexual with multiple partners, IV drug users, and patient with hemophilia. Most people are not aware of it at the time of infection and it may take 10 years or longer for AIDS to develop.
- **Herpes simplex infection**—Cold sores caused by herpes simplex infection are very infectious. There are chances of contracting herpetic whitlow, i.e. painful finger infection.
- **Tuberculosis**—it is caused by mycobacterium tuberculosis. If there is active infection, it may get transfer from saliva to the dentist.

Infection Risk Patients

- Patients who are HBs antigen positive and HBe antigen negative.
- Established cases of AIDS or AIDS related diseases like hairy leukoplakia, atypical ulceration and Kaposi's sarcoma.
- Patients who are HIV antibody positive.
- Patients who use IV drugs or other risk activities.

Methods of Decontamination

- **Sanitization**—this is first level of decontamination. This is process of physical cleaning to reduce quantity of microbes. This is done by cleaning the surface with soap and water or with detergent.
- **Disinfection**—this is next level of decontamination. It kills all vegetative microorganisms, fungi. This uses chemical germicides, radiation, ultraviolet rays or heat.
- **Sterilization**—this is third level. Sterilization kills all bacteria, fungi, virus and bacterial endospores. It uses chemical methods and physical methods.

Classification of Surface for Infection Control

According to Spaulding's classification in dental radiography is as follows:

- **Critical surface**—this is a surface that penetrates the tissue and contact blood. Examples are, scalpel blades, burs, extraction forceps, files, periodontal instruments, surgical drains, dental explorer, periodontal probes, biopsy punch, surgical drains, endodontic file reamer and implants. In this case, sterilization is done with the help of autoclave, chemical, dry heat.
- **Semi-critical surface**—this is the surface which does not penetrate tissue but contact with mucous membrane. It includes mouth mirrors, handpiece, anaesthetics syringe, chip syringes, amalgam condenser, impression trays, air/water syringe tips and high volume evacuator tips. In this case, high level disinfection and sterilization is required or barrier should be used.
- **Non-critical**—this includes the surface which penetrates neither tissue nor contact with mucous membrane. There is only contact with intact skin. It includes chair light handles, instrument trays, high touch work surface, bracket tales, chair control and dental chairs. In this case intermediated level of disinfection and sterilization is required. It is done by hydrogen peroxide bases, phenols, idophores.
- **Environmental surface**—this does not contact with patient directly. They can be contaminated by the care provider. It includes floor and door handles. In this case, sanitization should be properly done.

Working Area

The best method to reduce microorganism transfer is, to touch as few areas as possible.

- *Preparation of area*—the entire surface that is touched during radiography, should be covered with plastic wrap, plastic backed paper or aluminum foil. Covering eliminates the need for surface disinfectant after radiography.
- *After treatment*—if working area is not covered by protective covering, then after treatment of each patient, dental unit surface can become contaminated with blood and saliva. Operator should avoid touching walls and other surfaces with contaminated gloves. This area should be thoroughly cleaned with disposable towel.
- *Material used*—to clean working surface disinfecting solution is sprayed and allowed to dry. Most commonly used disinfectant is phenolic, idophores and chlorine containing compounds.

Sterilization

Sterilization is the process of destroying all the microbial life from an article or surface inclusive of bacterial spores. It can be achieved by:-

Dry Heat

It denatures the proteins of a microorganism rendering them non-viable. This principle is applied in the hot air oven. The instruments are packed loosely inside the oven and a temperature of 160°C is achieved with a holding time of 1 hour.

The hot air oven is exclusively used for sterilizing glass articles, oils, greases and powders. The other physical methods include microwave and ultraviolet methods (Fig. 6-2).

Moist Heat

Moist heat denatures and coagulates the proteins of a microorganism and is a better method of sterilization due to a higher efficiency of penetration than dry heat and a faster killing rate. This is due to latent heat of vaporization which is present in moist heat. This principle is used in the autoclave (Fig. 6-3).

Various methods of autoclaving are available base on the same principle. The temperature required for this is 121°C with a minimum holding time of 20 minutes. For practical consideration, vacuum models are operated at a temperature of 139°C, 332lb pressure and holding time of 5 minutes.



Fig. 6-2: Ultraviolet sterilizer.



Fig. 6-3: Autoclave used in dentistry.

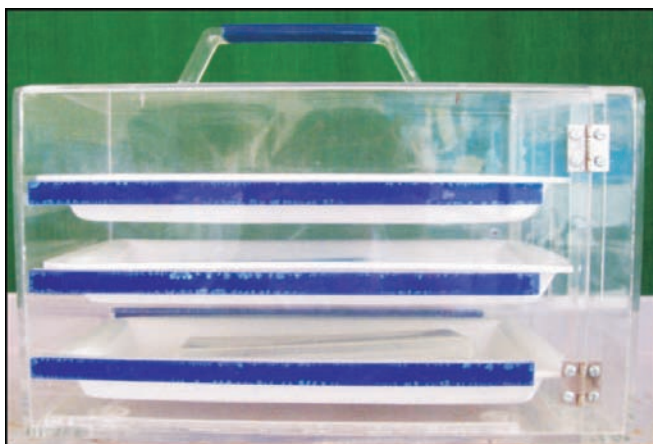


Fig. 6-4: Formalin chamber.

For gravity displacement models, a temperature of 121°C, 20lb pressure and a holding time of 30 minutes are used.

Root canal instruments and burs can be washed and thereafter sterilized in a glass bead sterilizer at a temperature of 210-230°C. The specific disadvantage of this sterilizer is that the handle portion is not sterilized and therefore, these articles are not entirely sterile.

Chemical (Fig. 6-4)

Ethylene oxide—this is a toxic gas that can be used to sterilize heat sensitive equipments. The articles are to be packed dry prior to sterilization otherwise a thin film of toxic ethylene glycol is formed. After sterilization, the articles should be aerated for at least 24 hours before use for dissipation of residue of the gas which could cause haematological complications. These are suited for large institutions and may not be a feasible option where small establishments are involved.

Glutaraldehyde—it is being marketed as a 2% solution which has to be activated with an alkali which is provided with the product as an activator, as only the alkaline solution is effective. The solution should have a contact time of at least 6 hours. A lesser contact time is employed for disinfection will not be effective against spores.

Disinfection

This is the process by which pathogenic microorganisms are removed from the surface without removing bacterial spores. This process is used to treat articles which do not penetrate mucous membrane or skin. Examples of disinfectants are formaldehyde, halogens, phenols and chlorhexidine.

Infection Control Procedures

The basis for infection control lies in the fact that, as dentist are more prone to acquire many infectious diseases; including blood borne diseases such as AIDS and hepatitis B. The procedure for infection control can be grouped into six major areas:

Training

All the staff members should be given training of infection control procedure. They should be told about the hazards due to infection which can occur to them or get transferred from them to patients. Continued education programme about infection control measures should be held regularly.

Vaccination

This is very important in infection control procedure. All clinical staff should be vaccinated against hepatitis B, etc. and regularly get it checked.

Covering of Wound

Open wound on the hand should be covered with waterproof dressing. Hand injuries during dental procedure should be avoided.

Hand Washing

It provides protection to both the patients and the dental team. Hand washing continues to be an important means for personal protection and of disease prevention. Washing hands before gloving reduces the skin microbial flora and helps to prevent skin irritation by waste products of bacterial growth under gloves.

Gloved hands should be washed under running water before and after taking radiograph of patient and a disinfectant like povidone iodine and 7.5% surgical scrubs or chlorhexidine gluconate 4% surgical scrub should be used.

Hand washing should be done at the start of treatment and after removing the gloves. Hand washing after using gloves is required to reduce the bacterial count that build on the skin during glove used. Specific step should be taken to hand wash. They are as follows (Fig. 6-5).

- Wet hands with warm water.
- Dispense an adequate amount of soap.
- Thoroughly rub the hands including area around the thumb and finger for 30 seconds.
- Wash hand with warm water to remove the soap.
- Dry hands with proper towel.



Fig. 6-5: Hand should be properly washed before gloving.

Gloves

Gloves used for patient care protect the dental team members from direct contact with patient microbes and they protect patients from contact with microbes on the hands of dental team member. Gloves need to be changed between patients and are to be washed with detergent. Gloves should be inspected periodically during patient care and torn or punched gloves should be removed as soon as possible and the hands should be washed.

Although some of these types, i.e. utility gloves can be autoclaved (nitrile type) through washing with an antimicrobial hand washing agent followed by rinsing and towel drying is adequate between routine uses.

Exam gloves are made from latex, vinyl, nitrile and polyurethane. While wearing the gloves, you should look for breaches in the integrity of gloves. Gloves should never be washed before use or disinfected for reuse. Grubby gloves and cleaning swabs should be placed in suitable disposable bags and sealed for incineration.



Fig. 6-6: Doctor should always wear double gloves while examination of the patient.

Protection against Aerosol and Spatter

It involves the use of a pre-procedural rinsing of the patient's mouth with an anti-microbial solution, use of rubber dam, high volume evacuation, protective clothing and eyewear.

Reduce the amount of spray and spatter that exist in a patient's mouth during care, which in turn reduce the number of microbes contaminating the dental team.

Rubber dam (Fig. 6-7) has been shown to reduce the number of microbes in dental aerosols generated during operative procedures and saliva ejection minimizes saliva form accumulating in the mouth.



Fig. 6-7: Rubber dam should be used to control dental aerosols generated during operative procedure.

Having patient rinse their mouth with an antimicrobial mouth rinse just before care, will reduce the number of bacteria that exist in the mouth.

Eyewear and face shield—eyewear and face shields with adequate side protection prevent spatter from patient's mouth or splashes of contaminated solution and chemicals from contacting eyes (Figs 6-8 and 6-9).



Fig. 6-8: Dental professional should wore eyewear to prevent spatter from patient mouth.

Masks—face masks prevent spatter from patient's mouth or splashes of contaminated solution and chemicals from contacting the mucous membrane of mouth and nose. A secondary protective aspect of a mask is the reduction in the inhalation of airborne particles. Thus, masks should be worn whenever there is a risk of aerosol generation, spraying, spattering, or splashing of patient's oral fluid or chemicals used (**Fig. 6-10**).

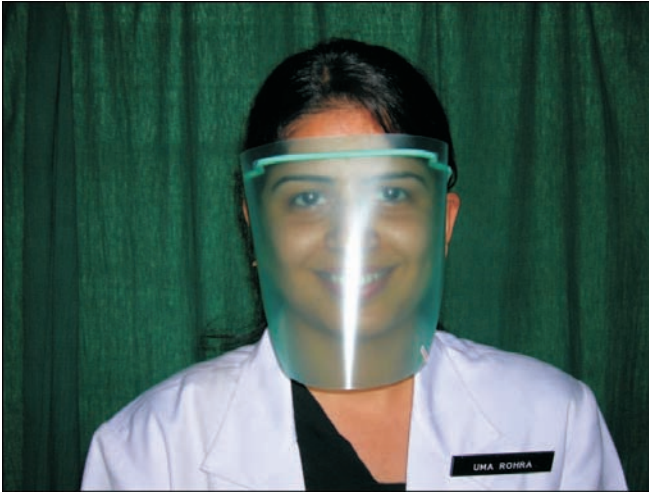


Fig. 6-9: Complete face shield also help to protect from aerosols from patient oral cavity.

Protective clothing—as with eyewear and masks, protective clothing is also worn under conditions generating sprays, spatters, splashes or spills of body fluids, contaminated solutions. Protective clothing is the outer layer covering the garment that would first be contacted by the contaminating droplets. Such fluid resistant garments for use at chair side in dentistry may be reusable or disposable but it should protect the skin and underlying clothes from



Fig. 6-10: While examining the patient, dental professional should wear mask, gloves.

exposure to potentially infectious material. This protection can be provided by high-neck, long sleeved, knee length garments.

Instrument Asepsis

You should provide instruments that are safe for patient's use. Pre-soaking of contaminated instruments keeps them wet until thorough cleaning can occur. This procedure prevents blood and saliva from drying on the instrument which facilitate cleaning.

A cleaning solution manufactured for use in ultrasonic cleaners should be used and it needs to be changed daily. Protection against splashing and direct contact with this contaminated solution is important. Usually 2 to 20 minutes are needed to clean instruments ultrasonically as determined by observing the cleanliness of the processed instruments and making appropriate adjustments.

After cleaned instruments have been rinsed and dried, they are to be packaged in functional sets before sterilization. Instrument packages sterilized in steam becomes wet and must be allowed to dry before handling so that the packing does not tear.

Surface Asepsis

It eliminates the involvement of surface in the spread of diseased agents.

There are two general approaches to surface asepsis. One is to clean and disinfect contaminated surface and the other is to prevent the surface from becoming contaminated by use of surface covers.

Disinfection of surfaces involves the cleaning of surface after every patient and application of a disinfectant chemical. These chemicals include alcohol, phenols, glutaraldehyde, chlorine etc.

Spittoons should be flushed with water.

Management of Sharp and Other Waste Products

It reduces the chances for sharp injuries and contact with potentially infectious materials. There should be safe handling of needles and safe removal of needles. Needles should not be recapped instead, should be disposed off directly. Handpiece should not be left in position with burs; instead the bur should be removed after use. Infectious waste is preferably done by incineration.

Aseptic Technique

It includes aseptic retrieval of supplies, reducing contamination from dental unit water supplies, aseptic radiographic procedures.

Suggested Reading

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Section **2**

Diagnostic Procedures



7

Case History

Introduction

A case history can be considered to be a planned professional conversation that enables the patient to communicate his symptoms, feeling and fears to the clinician so that the nature of the patient's real and suspected illness and mental attitudes may be determined.

The interest, warmth and compassion exhibited by the dentist are important factors in establishing patient rapport and in obtaining meaningful history. A kind and considerate approach is most important in securing and gaining the confidence of the patient. Indicate the patient that you are a friend who is keenly interested in him/her as a person.

Ideally the patient's history should be taken in a consultation room or a private office in which the décor and the furnishing are quite different from those of the dental operatory. The friendly atmosphere is an important factor in helping the patient to talk freely about his problems (Fig. 7-1).



Fig. 7-1: Taking and recording history of the patient is important part in diagnostic sequence.

Diagnostic Procedure (Fig. 7-2)

The procedure can be divided into:-

- Personal information.
- Taking and recording history.
- Examining the patient.
- Establishing a provisional diagnosis on the basis of history and examination.
- Conducting the necessary investigation.
- Formulation of final diagnosis on the basis of the results of the investigations.
- Making a plan of treatment and medical risk assessment for dental patient

Personal Information

Name

A patient usually likes to be called by name. This will help to elicit the history properly, but it will also be of psychological benefit to the patients. In case of pediatric patients, addressing the patient by his/her name or pet name will encourage him/her to talk freely. Advantage of knowing the patient names are identification, to maintain record, communication and psychological benefit

Age

Knowing the patient's age is beneficial to the clinician in more ways than one.

Diagnosis

Certain diseases are more common at certain ages (Tables 7-1 to 7-4).

Treatment Planning

- *Absence of teeth*—if there is complete absence of teeth (i.e. true anodontia) even at the age of 4-5 years; it is most

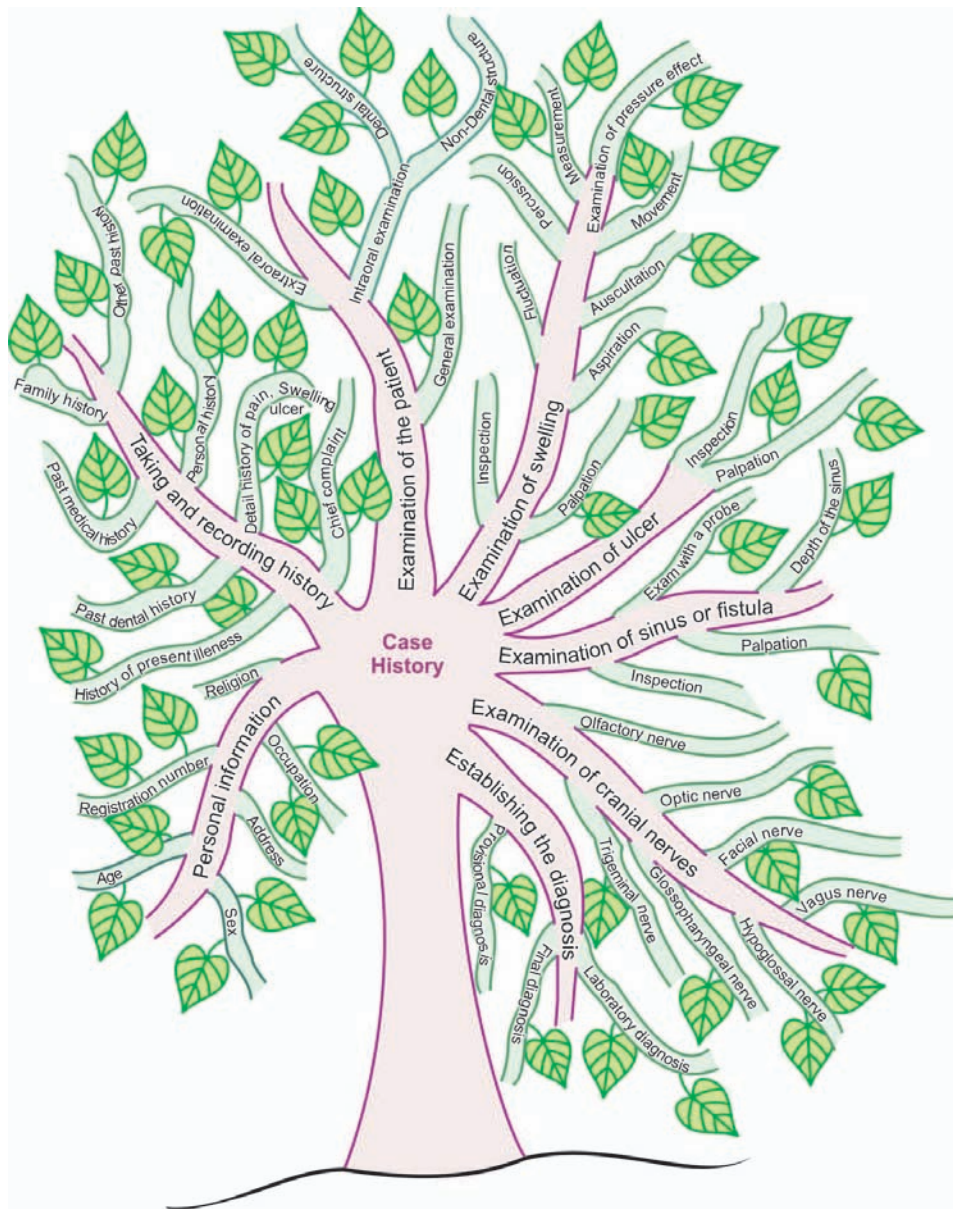


Fig. 7-2: Diagrammatic representation of diagnostic procedure of case history.

frequently in association with hereditary ectodermal dysplasia.

- **Delayed eruption**—delayed eruption of teeth may be associated with rickets, cretinism or certain local factors such as fibromatous gingivae. If the permanent tooth does not erupt in its eruption period, we have to check by radiograph whether the permanent tooth bud is present or not. If the bud is present, deciduous tooth should be extracted.
- **Ankylosed deciduous teeth**—ankylosed deciduous teeth prevent the eruption of permanent teeth; in such cases,

ankylosed teeth should be removed surgically to prevent the development of malocclusion, local periodontal disturbances and dental caries.

- **Behavior management technique**—it differs according to the age of the patient. In case of pediatric patients, the dentist has to deal with the child as well as with the parent; hence in pediatric dentistry, the approach is 1:2; while in case of adults, the approach is 1:1. In talking to a child, the dentist must get down to the patient's own level, in position and in conversation. Child patients may be intelligent or retarded.

Table 7-1: Disease present at/since birth

DENTAL DISEASES**Related to jaw**

- Agnathia
- Facial hemihypertrophy
- Macrognathia
- Cleft palate
- Facial hemiatrophy

Related to lip

- Commissural pits and fistulae
- Double lip
- Cleft lip
- Hereditary intestinal polyposis syndrome

Related to gingiva

- Congenital epulis of the newborn
- Fibromatosis gingiva

Related to rest of oral mucosa

- Pigmented cellular nevus
- Cystic hygroma
- Fordyce's granule

Related to tongue

- Microglossia
- Macroglossia
- Aglossia
- Ankyloglossia
- Cleft tongue
- Fissured tongue
- Median rhomboidal glossitis
- Lingual thyroid nodule

Related to salivary glands

- Aplasia
- Atresia
- Aberrancy
- Developmental lingual salivary gland depression

Related to teeth

- Pre-decidual dentition

Related to TMJ

- Aplasia or congenital hypoplasia of the mandibular condyle

Others

- Teratoma
- Erythroblastosis fetalis
- Hemophilia

SYSTEMIC DISEASES**Related to heart**

- Atrial septal defect
- Patent ductus arteriosus
- Ventricular septal defect
- Pulmonary stenosis
- Aortic stenosis
- Tetralogy of Fallot

Related to respiratory system

- Bronchiolitis
- Cystic fibrosis

Related to CNS

- Congenital myopathy
- Aqueduct stenosis

Related to alimentary tract

- Meckel's diverticulum (it is common congenital anomaly. It is a small hollow wide mouthed sac protruding from ileum)
- Annular pancreas (pancreas encircle the 2nd and 3rd part of duodenum leading to gastric outlet obstruction)

Table 7-2: Diseases more commonly seen in infancy

Dental/oral/systemic diseases with oral manifestations

- Palatal cyst of the newborn
- Heterotrophic oral gastrointestinal cyst
- Hemangioma
- Fibrosarcoma
- Hemangioendothelioma
- Hemangiopericytoma
- Embryonic rhabdomyosarcoma
- Melanotic ameloblastoma
- Dental lamina cyst of the newborn
- Hurler's syndrome
- Acute suppurative osteomyelitis of the jaw
- Letterer-Siwe disease
- Progeria
- Infantile cortical hyperostosis of the jaw
- Fibrous dysplasia of the jaw
- Sprue
- Thalassemia

Systemic diseases

- Congenital heart diseases
- Bronchiectasis
- Pneumonia

- *Child dose*

$$\text{Young rule} = \frac{\text{Child's age}}{\text{Age} + 12} \times \text{adult dose}$$

$$\text{Clark rule} = \frac{\text{Child age at next birth day}}{24} \times \text{adult dose}$$

$$\text{Dilling rule} = \frac{\text{Age}}{20} \times \text{adult dose}$$

Sex

- *Diagnosis*—certain diseases are more common in a certain sex (**Tables 7-5 and 7-6**).
- *Gifting*—in case of pediatric patients, gifting varies according to the sex of the child.
- *Esthetics*—females are much conscious about esthetics.
- *Emotion*—females are emotional and sensitive hence one must be very careful during treatment. Failure to thrive, educational and emotional abuse are most common in case of female patients.
- *Child abuse*—sexual abuse or exploitation is more common in case of females.
- *Dose*—females have smaller body weight and require lower dose as compare to males.
- *Drug interaction*—in females, consideration must be given to menstruation, pregnancy and lactation. Drugs given during pregnancy can affect the fetus. Antihypertensive drugs interfere with sexual function in males but not in females. Gynecomastia is a side effect of ketoconazole, metronidazole, chlorpromazine and digitalis administrations that occur in males. Ketoconazole causes loss of libido in men but not in women.

Table 7-3: Diseases frequently seen in children and young adults**Dental/oral/systemic diseases with oral manifestations**

- Focal epithelial hyperplasia
- Retrocuspid papilla
- Fissured tongue
- Benign migratory glossitis
- Benign cervical lymphoepithelial cyst
- Epidermoid and dermoid cyst
- Papilloma
- Papillon-Lefèvre syndrome
- Peripheral ossifying fibroma
- Juvenile periodontitis
- Naso-alveolar cyst (12-75 years)
- Thyroglossal duct cyst
- Oral submucous fibrosis (20-40 years)
- Osteoid osteoma of the jaw
- Benign osteoblastoma of the jaw
- Torus palatinus
- Kaposi's sarcoma
- Ewing's sarcoma
- Osteosarcoma of the jaw
- Primary lymphoma of bone
- Burkitt's lymphoma
- Hodgkin's disease
- Rhabdomyosarcoma
- Alveolar (soft part) sarcoma
- Pleomorphic adenoma
- Central odontogenic fibroma
- Odontogenic myxoma (2nd and 3rd decade)
- Peripheral cemental dysplasia
- Benign cementoblastoma
- Dentinoma
- Ameloblastic fibro-odontoma
- Reiter's syndrome (20-30 years)
- Chronic focal sclerosing osteomyelitis of the jaw
- Chronic osteomyelitis with proliferative periostitis of the jaw
- Basal cell carcinoma
- Lymphoepithelial cyst
- Lingual varices
- Sq. cell carcinoma
- Scarlet fever
- Diphtheria
- Sarcoidosis
- Nasopharyngeal angiofibroma (16 years)
- Chondromyxoid fibroma (under 25 years)
- Benign chondroblastoma of the jaw (5 to 25 years)
- Osteoma of the jaw (young adult)
- Rickets
- Sickle cell anemia
- Infectious mononucleosis
- Acute leukemia
- Pemphigus
- Noma
- Primary herpetic stomatitis
- Recurrent aphthous stomatitis
- Acute lymphonodular pharyngitis
- Dental caries
- Nursing bottle caries (children)
- Pulp polyp
- Eruption cyst
- Dentigerous cyst (2nd decade)

Systemic diseases

- | | |
|-----------------------------|-----------------------------|
| • Congenital heart diseases | • Rheumatoid heart diseases |
| • Asthma | • Acquired bronchiectasis |
| • Pneumonia | • Jaundice |
| • Smallpox | • Juvenile diabetes |
| • Typhoid | • Cretinism |

Table 7-4: Diseases frequently seen in adults and older patients**Dental/oral/systemic disease with oral manifestation**

- | | |
|--|--|
| • Attrition | • Abrasion |
| • Gingival recession | • Periodontitis |
| • Acute necrotizing ulcerative gingivitis | • Dentinal sclerosis |
| • Pulp stone and fibrosis | • Cementicles |
| • Root resorption | • Atrophic diffuse sclerosing osteomyelitis of the jaw |
| • Lichen planus | • Leukoplakia |
| • Leukoedema | • Erythroplakia |
| • Carcinoma <i>in situ</i> | • Sjogren's syndrome (over 40 years) |
| • Adenoid cystic carcinoma | • Mucoepidermoid carcinoma |
| • Acinic cell carcinoma | • Gingival cyst of the adult |
| • Necrotizing sialometaplasia | • Primary intra-osseous carcinoma of the jaw |
| • Ameloblastoma (30-50 years) | • Macroglobulinemia |
| • Adenoameloblastoma | • Ameloblastic fibrosarcoma |
| • Sq. odontogenic tumor | • Carcinoma of gingiva |
| • Peripheral odontogenic fibroma | • Spindle cell carcinoma |
| • Trigeminal neuralgia | • Orolingual paresthesia |
| • Verrucous carcinoma | • Glossopharyngeal neuralgia |
| • Adenoid sq. cell carcinoma | • Malignant melanoma |
| • Sphenopalatine neuralgia | • Peripheral giant cell granuloma (30 years) |
| • Temporal arteritis | • Median anterior maxillary cyst (40-60 years) |
| • Fibroma | • Nasoalveolar cyst |
| • Carcinoma of lip, tongue, floor of mouth and buccal mucosa | • Lingual cyst (7-99 years) |
| • Keratoacanthoma (50-70 years) | • Herpes zoster |
| • Lymphoepithelial cyst (15-65 years) | • Pernicious anemia |
| • Basal cell carcinoma | • Osteomalacia |
| • Chronic leukemia | • Canicular adenoma |
| • Iron deficiency anemia | • Adenolymphoma |
| • Pleomorphic adenoma | • Hereditary hemorrhagic telangiectasia |
| • Oxyphilic adenoma | • Osteoma of jaw bone |
| • Lipoma | • Fibrosarcoma of jaw bone |
| • Myxoma of jaw bone | • Hodgkin's disease |
| • Chondroma of jaw bone | • Solitary plasma cell myeloma |
| • Torus mandibularis | • Malignant schwannoma |
| • Liposarcoma of jaw bone | |
| • Multiple myeloma | |
| • Leiomyoma | |
| • Olfactory neuroblastoma | |

Systemic diseases

- | | |
|--|--|
| • Diabetes | • Hypertension |
| • Hypotension | • Congestive cardiac failure and myocardial infarction |
| • Renal failure | • Chronic valvular heart disease |
| • Atherosclerotic diseases like angina pectoris and coronary heart disease | • Hepatitis |
| • Peptic ulcer | • Asthma |

Table 7-5: Diseases more common in females

- | | |
|--------------------------------|---------------------------------------|
| • Iron deficiency anemia | • Caries |
| • Diseases of thyroid | • Pleomorphic adenoma |
| • Sjogren's syndrome | • Adenoameloblastoma |
| • Central cementifying fibroma | • Myasthenia gravis |
| • Torus palatinus | • Peripheral cemental dysplasia |
| • Sickle cell anemia | • Juvenile periodontitis |
| • Cicatricial pemphigoid | • Recurrent aphthous stomatitis |
| • Malignant melanoma | • Peripheral giant cell granuloma |
| • Central giant cell granuloma | • Peripheral ossifying fibroma |
| • Nasoalveolar cyst | • Idiopathic thrombocytopenic purpura |

Table 7-6: Diseases more common in males

• Attrition	• Caries in deciduous teeth
• Carcinoma <i>in situ</i>	• Carcinoma of the buccal mucosa
• Keratoacanthoma	• Basal cell carcinoma
• Leukoplakia	• Adenoid cystic sq. cell carcinoma
• Verrucous carcinoma	• Multiple myeloma
• Liposarcoma	• Chondrosarcoma
• Hodgkin's disease	• Herpes simplex
• Solitary plasma cell myeloma	• Ewing's sarcoma
• Wegener's granulomatosis	• Ameloblastic fibro-odontoma
• Osteoid fibroma	• Basal cell adenoma
• Benign osteoblastoma	• North American blastomycosis
• Reiter's syndrome	
• Adenolymphoma	
• Pernicious anemia	

Address (Residence)

- **Correspondence**—it is necessary for future correspondence. Full postal address of the patient should be taken for future communication.
- **Geographical prevalence of dental/oral diseases**—a few diseases have got geographical distribution. *Dental caries* and *mottled enamel* are dependent on the fluoride content of the domestic water. In India, *cancer of the lip* is very rare and *cancer of tongue* and *buccal mucosa* constitute the bulk of oral cancers. In cancer patients from *Mumbai*, common site affected is the tongue and in *Chennai*, it is the buccal mucosa. *Dental caries* is more common in modern industrialized areas while *periodontal diseases* are more common in rural areas. Habits also vary according to residence. Chutta, a form of tobacco, is common in *Tamilnadu*, gutkha in *Bihar*, hukli in *Bhavnagar district of Gujarat*. Bajjar is a dry snuff used by women in *Gujarat*.
- **Geographical prevalence of medical disease**—there are many medical diseases which have got geographical prevalence. For example *filariasis* (*Orissa*), *leprosy* (*Bankura district of West Bengal*), *gall bladder disease* (*West Bengal and Bangladesh*), *peptic ulcer* (*North Western and Southern parts of India*), *sleeping sickness* (*South Africa*), *hydatid disease* (*Australia*), *tropical disease* (*tropical countries*), *kangri cancer* (*Kashmir*), *anemia and malnourishment* (*rural areas*) *obesity, arthrosclerosis* (*urban area*) and *other industry related diseases* (*urban areas*).

Registration Number

It is good to give each and every patient a unique registration number and to maintain his/her records under that number so that when the patient visits the doctor at a later date, the doctor can know the details of the patient and the treatment done before.

Occupation

- **Financial status**—treatment varies according to the financial status of the patient. Expensive treatment

cannot be given to the patient with low socio-economic status.

- **Disease**—some diseases are peculiar to certain occupations:
 - **Attrition**—Certain occupations, in which the worker is exposed to an atmosphere of abrasive dust and cannot avoid getting the material into his mouth, causes severe attrition. Habitual opening of the pins may result in notching of the incisal edge.
 - **Abrasion**—it is commonly noted in carpenters, shoemakers or tailors who hold pins, nails or tacks between their teeth.
 - **Gingival staining**—the strange dark stippling of the marginal gingiva is seen in patient works with lead, bismuth or cadmium.
 - **Erosion**—undue erosion of teeth is seen in sandblaster, who should be promptly referred to his physician for pulmonary silicosis.
 - **Hepatitis-B**—dentists, surgeons, blood bank personnel etc. are very prone to hepatitis -B.
 - **Chelitis glandularis**—in occupations where there is chronic exposure to sun, wind and dust can cause cheilitis glandularis.
 - **Varicose veins**—it is common in bus conductors and traffic policemen.
 - **Urinary bladder neoplasm**—it is common in aniline dye workers.
 - **Carcinoma of scrotum**—it is common in chimney sweepers or those who work in tar and oil companies.
 - **Medial semilunar cartilage injury**—injury to the medial semilunar cartilage of the knee occurs in football players and miners.
 - **Student elbow**—enlargement of certain bursae may occur from repeated friction of the skin over the bursae like in 'student's elbow', 'tennis elbow' and 'house-maid knee'.
 - **Mesothelioma**—an ice cream vendor may have mesothelioma due to exposure to asbestos for many years.
 - **Stress disease**—certain occupational hazards are associated with office work, e.g. repetitive strain injury and migraine induced by stress of inappropriate lighting.
 - **Cirrhosis of liver**—barmen who have ready access to alcohol have high mortality for cirrhosis of liver and tuberculosis.
 - **Countryman's lip**—carcinoma of lip is commonly seen in persons who have to do outdoor work and that's why, it is called as 'countryman's lip'
- **Time**—In case of pediatric patients, if both the parents are working and even if they are aware of the importance of dental health, they cannot properly attend the appointments given by the dentist.

Religion

- *Carcinoma of penis*—it is hardly seen in Jews and Muslims owing to their religious custom of compulsory circumcisions of penis.
- *Phimosis*—for the same reason phimosis, subprepuical infection etc are not seen in them.
- *Intussusception*—on the other hand intussusception is sometimes seen after the month long fast in Ramjan in Muslims.

Taking and Recording History

Chief Complaint

- *Definition*—it is the patient's response to the dentist's questions. Chief complaint is the reason for which the patient has come to doctor or the reason for seeking the treatment. The chief complaint should be recorded in the patient's own words and first attention should be given to it.
- *Chronological recording*—each of these complaints should be recorded in chronological order. If few complaints start simultaneously, record them in the order of severity.
- *Do not interrupt*—do not interrupt the patient if possible.
- *Significance*—chief complaint aids in the diagnosis and treatment planning.
- Most common chief complaints and their causes are described in **Table 7-7**.

History of Present Illness

- *Collecting information*—the history commences from the beginning of the first symptom and extends to the time of examination. Information should be collected by asking the following questions:
 - When did the problem start?
 - What did you notice first?
 - Did you have any problems or symptoms related to this?
 - Did the symptoms get better or worse at any time?
 - What you done to treat these symptoms?
 - Have you consulted anyone else for this problem?
- *Mode of onset*—it can be sudden or gradual. It should be in terms of time, in days, weeks, months, before the current appointment.
- *Cause of onset*—one has to determine whether if any thing that the patient has experienced may have precipitated the signs and symptoms.
- *Duration*—it should be noted that, since how many days complaint was present.
- *Progress*—it may be described as intermittent, recurrent, constant, increasing or decreasing in severity. Aggravating and alleviating factors should be noted.

- *Relapse and remission*—this should be noted. If patient has got any similar complaint in the past and then again it relapse.
- *Treatment*—if the patient has taken any treatment for the same problems, the mode of treatment and doctor who has treated the patient should be recorded.
- *Negative history*—e.g. in case of sinus on the cheek absence of the history of watery discharge at the time of meals excludes the possibility of the parotid fistula.

History with Particular Reference

Pain (Fig. 7-3)

- *Anatomical location where the pain is felt*—the patient should be asked to point to the area affected and outline the way in which the pain may spread to affect other areas. He should be asked whether the pain is deep or superficial.
- *Origin and mode of onset*—the onset may be wholly spontaneous. It may be induced by certain activities like yawning, chewing, drinking hot and cold liquids etc. It may be triggered by minor stimulation such as touch or movement of skin, lip and tongue.

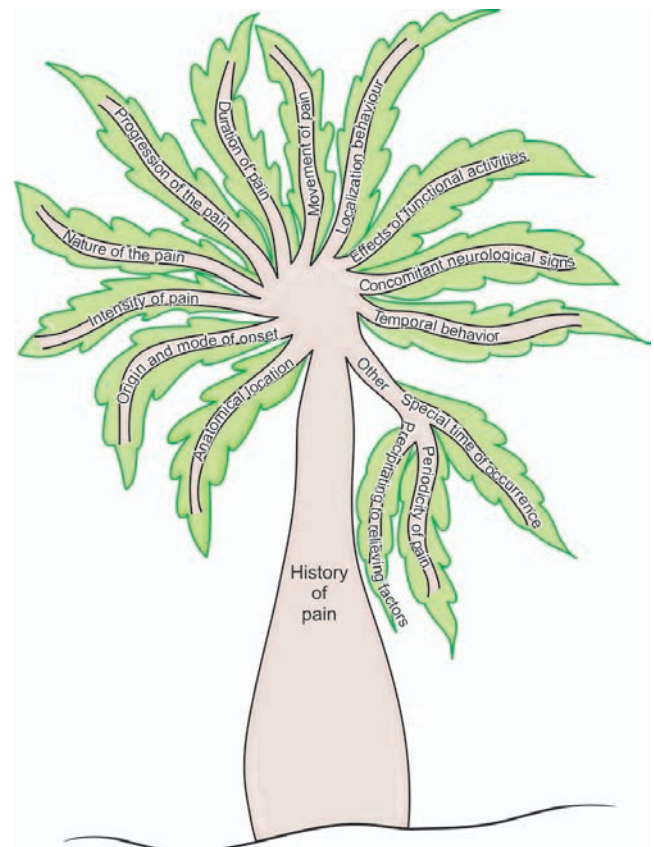


Fig. 7-3: History of the patient in particular reference to pain.

Table 7-7: Most common complaints and their causes

Pain - Gingival and periodontal disease - Pulpal disease - Mucous membrane disease - Salivary gland infection - Lesions of jaw bones - Lymph node inflammation - TMJ disorder - Maxillary sinus disease - Ear disease - Psychosis - Angina pectoris - Tonsillar disease - CNS disease - Neuralgia - Neuritis - Esophageal diverticulum - Eagle's syndrome - Trotter's syndrome	- Trauma - Normal resorption of deciduous teeth - Pulpoperiodontal disease - Malignant tumor - Chondroma - Myxoma - Hemangioma - Histiocytosis X - Familial hypophosphatemia - Papillon-Lefèvre syndrome - AIDS	Bad taste - Aging changes - Heavy smoking - Poor oral hygiene - Dental caries - Periodontal disease - ANUG
Burning sensation - Idiopathic burning tongue - Psychosis - Neurosis - Viral infection - Fungal infection - Chronic bacterial infection - Geographic tongue - Xerostomic condition - Fissured tongue - Generalized oral mucosal disease - Anemia - Achlorhydria - Multiple sclerosis - Vitamin deficiency	Recent occlusal problems - Periodontal disease - Traumatic injury - Periapical abscess - Cyst and tumor of tooth bearing region of jaw - Fibrous dysplasia	Diabetes - Hypertension - Medication - Psychosis - Neurological disorder - Decreased salivary flow - Uremia - Intraoral malignancy
Bleeding - Gingivitis - Periodontal disease - Inflammatory hyperplasia - Allergy - Traumatized tumor - Hemangioma - Traumatic injury - Deficiency of coagulation factors	Delayed tooth eruption - Malposed or impacted teeth - Cyst - Odontoma - Sclerosed bone - Tumor - Mal-development - Cleidocranial dysplasia - Hypothyroidism	Paresthesia and anesthesia - Injury to regional nerve—anesthetic needle and jaw bone fracture - Malignancy - Medications like those used in sedation, hypnosis - Diabetes - Pernicious anemia - Multiple sclerosis - Acute infection of jaw bones - Psychosis
Loose teeth Periodontal disease	Xerostomia - Local inflammation - Infection and fibrosis of major salivary gland - Dehydration state - Drugs like tranquilizers and antihistaminic - Autoimmune diseases like Sjogren's syndrome and Mikulicz's disease - Chemotherapy - Post radiation changes - Psychosis Swelling - Inflammation and infection - Cyst - Retention phenomenon - Inflammatory hyperplasia - Benign tumor - Malignant tumor	Halitosis - Poor oral hygiene - Periodontal disease - Third molar opercula - Decayed tooth - ANUG - Oral cancer - Spicy foods - Tobacco use - Nasal infection - Tonsillitis - Pharyngeal infection and tumor - Gastric problems - Diabetes - Uremia

- Intensity of pain**—the intensity of pain should be established by differentiating between mild and severe pain. This will show how patient reacts to his suffering. Intensity can be recorded verbally or by some scaling method.
- Nature of the pain**—the general quality of the pain complaint should be classified as how it makes the patient feel. It can be burning, throbbing, scalding, shooting, stabbing, vague aching pain, splitting, dull, aching, lacerating, cutting, boring, constricting, gripping, scorching, pounding, heavy, pressing, sharp, bright, gnawing, cramping, squeezing, or searing.
- Progression of the pain**—the progression of pain should be noted.
- Duration of pain**—the duration of an individual pain episode is an important descriptive feature that aids in pain identification. A pain is said to be momentary if its duration is expressed in seconds. The pain that continues from one day to the next is said to be protracted.
- Movement of pain**—radiation of pain, referred pain and shifting or migration of pain.
- Localization behavior**—a determination of pain localization should be made in the description of the behavior. This includes whether the pain is localized or diffuse, radiating or spreading and enlarging or migrating.
- Effect of functional activities**—the effect of various activities such as talking, brushing of teeth, shaving, washing the face, turning the head, lying down, etc. should be noted.

The effect of tension, fatigue and the time of the day should also be noted.

- **Concomitant neurological signs**—the description of pain cannot be completed unless other sensory, motor and autonomic effects are included. Accompanying sensation of hyperesthesia, anesthesia or dysesthesia should be mentioned. Any concomitant change in the special senses affecting vision, hearing, smell or taste should be noted. Motor changes such as muscular weakness, muscular contraction should be described; likewise ocular signs, nasal signs, and cutaneous signs should be described.
- **Temporal behavior**—the temporal behavior of pain should be evaluated, whether the pain is intermittent or continuous. It should not be confused with the effect of medication that induces a period of comfort by analgesic action.
- **Others**—other point like special time of occurrence, periodicity of pain and precipitating factors and relieving factors should be noted.
- **Questions to be asked to the patient**—
 - Is the pain sharp and lacinating or dull aching and throbbing?
 - Does pain occur spontaneously or do certain activities cause it?
 - Is the pain constant or periodic?
 - Does the pain gradually build in intensity or it is paroxysmal?
 - Is the pain worse at any time of the day?
 - Is the pain of short duration or long?

Swelling (Fig. 7-4)

- **Duration**—since how many days swelling is present should be noted as it will determine its acute or chronic nature.
- **Mode of onset**
 - *Masses that increase in size just before eating*—salivary gland retention phenomenon.
 - *Slow growth*—reactive hyperplasia, chronic infection, cysts and benign tumors.
 - *Moderately rapid growing mass* (week to about 2 months)—chronic infection, cysts and malignancy.
 - *Rapidly growing mass*—abscess, infected cyst, aneurysm, salivary retention phenomenon and hematoma.
 - *Mass with accompanying fever*—infection and lymphoma.
- **Symptoms**—symptoms associated with swelling like pain, difficulty in respiration, swallowing or any other movement; disfiguring etc. In case of pain; nature, site and time of onset should be noted.
- **Progress of swelling**—swelling can increase gradually in size or rapidly.

- **Associated features**—fever, presence of other swellings and loss of body weight should be asked and noted.
- **Secondary changes**—like softening, ulceration, inflammatory changes.
- **Impairment of function**—nature of loss of movement and intensity.
- **Recurrence of the swelling**—if the swelling recurs after removal, it may indicate malignant changes. Cystic swelling may recur if the cyst wall is not completely removed.

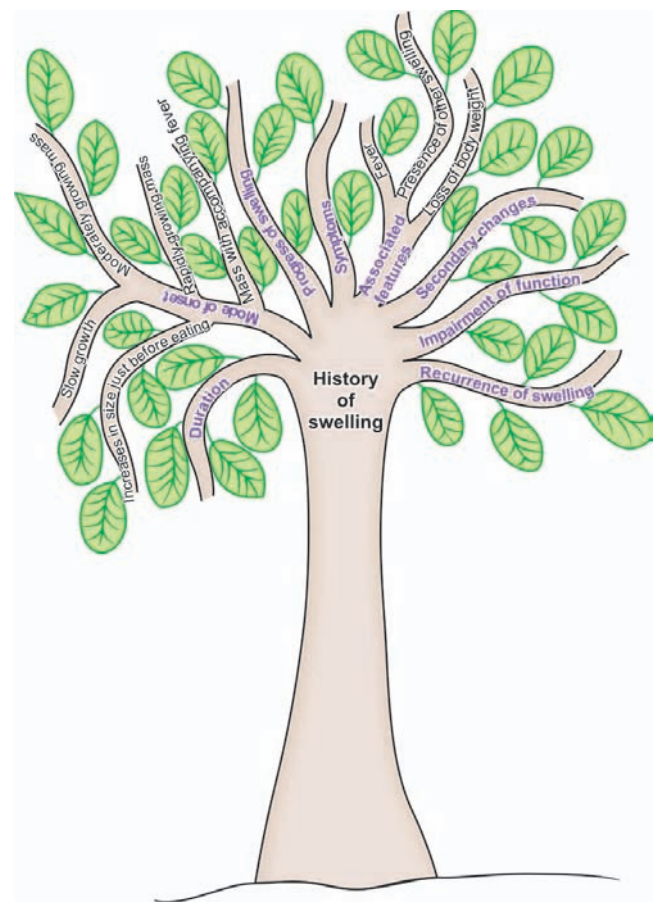


Fig. 7.4: History of swelling should be taken properly.

Ulcer

- **Mode of onset**—mode of onset and duration of the ulcer should be asked and recorded.
- **Pain**—ulcers associated with inflammation are painful; ulcers associated with epithelial or basal cell carcinoma are painless.
- **Discharge**—serum, blood and pus.
- **Associated diseases**—tuberculosis, nephritis, diabetes, and syphilis.

Past Dental History

- *Patient attitude*—in addition to chief complaint and history of the present illness, it is necessary to elicit a past dental history. It is necessary to obtain description of the details of previous dental treatment and his reaction to his dentist and the treatment. By noting this, you may get accurate idea of the importance he gives to good dental treatment and how conscious he has been in pursuing a goal of good oral health.
- *Component of past dental history*—significant component of past dental history include previous restorative, periodontic, endodontic or oral surgical treatments, reasons for loss of teeth, untoward complication of dental treatment, experience with orthodontic appliance and dental prostheses; and radiation or other treatment for past oral and facial lesions.

Past Medical History

Obtaining the patient medical history is very important as it can assess the patient health status and also it can facilitate for the better diagnosis for the orofacial complaint of the patient. By taking proper medical history, following goal are achieved.

- *Assess in diagnosis of oral disease*—there are many systemic problems which have oral manifestation. So by taking medical history, we can come to diagnosis of many orofacial complaints. For example, in HSV infection, negative history of recurrent herpes labialis and positive history of close contact with patient with primary and recurrent herpes is helpful in making the diagnosis.
- *Detection of underlying systemic problems*—many times, patient is not aware of systemic problems due to his negligence. So by taking proper medical history, we can detect many systemic problems in the patient.
- *Management of patient*—by assessing the patient general status, we can determine our plan of treatment. As many systemic diseases can change our line of treatment while treating the dental complaint. So we can modify our treatment according to need.
- *Consultation with other professional*—by assessing the medical status, we can also determine whether any additional information is necessary to assess the patient medical status. Dentist may require consultation in following conditions:
 - *Known medical problems*—consultation is required in patients who have known medical problems and schedule for stressful dental procedure.
 - *Unknown medical problems*—in some patients abnormalities are detected while history taking or physical examination or laboratory studies. Patient is unaware of this problem.

- *High risk patient*—some patient have high risk for development of particular problems. For example, obese patient may have hypertension.
- *Additional information*—in patient who require additional information which may alter dental care and assist in the diagnosis of orofacial problems.

Consultation letter

Place

Date

Patient Name

Referral doctor name

To

Dr X

I am referring the patient ——— who has to undergo stressful procedure. We have to use local anesthesia for this patient. Medical history of patient reveals that he is suffering from ———. Please advice us about the status of the patient whether he can undergo stressful dental procedure, which type of local anesthesia should we use and what precaution should we take.

Patient signature

Date

Doctor signature

Date

Please return this consultation to

- *Medical questionnaire*—one should ask the following particulars to the patient.
 - *Systemic problems*—whether the patient was suffering from any medical problems before? If yes, what was the duration and treatment? Whether the treatment was beneficial or not? Whether he had taken any medication for that? Patient must be reminded that medicine may include anything taken into the body that is not food. All diseases suffered, by the patient, previous to the present one should be recorded. These diseases may not have any relation with present disease; particular attention must be given to diseases like diabetes, asthma, bleeding disorders, hypertension, myocardial infarction, hepatitis B, diphtheria, rheumatoid heart disease, tuberculosis, and gonorrhea.
 - *Chest pain*—history of chest pain should be asked. As we will come to know the cardiological status of the patient.
 - *Allergy*—whether he has any allergy? (According to that the treatment plan may be adjusted). Allergy can be due to any drug or food. Patient should be asked

about asthma, eczema and hay fever, urticaria, angioedema etc.

- *Previous hospitalization*—ask the patient to enumerate all previous hospitalizations and indicate the purpose.
- *Blood transfusion*—history of blood transfusion should be asked.
- *Accident and operation*—Previous operations, fractures, and accidents should also be noted.
- *Drug history*—you should ask patient to tell the medication that they are presently taking.

Personal History (Fig. 7-7)

- *Habits and addiction*—it is important aspect of history taking. There are many diseases which can correlate with particular habit of the patient. The most important habit you should ask about the tobacco, smoking and alcohol. You should note down frequency per day and length of time that patient had habit in years.
- *Oral hygiene and brushing technique*—bad oral hygiene and improper brushing technique may lead to dental caries and periodontal disease. Horizontal brushing technique may lead to cervical abrasion of the teeth.
- *Pressure habits*—thumb sucking, lip sucking, and finger sucking may lead to anterior proclination of maxillary anterior teeth.
- *Tongue thrusting*—it may lead to anterior and posterior open bite and proclination of anterior teeth.
- *Mouth breathing*—it may lead to anterior marginal gingivitis and caries (Fig. 7-5).



Fig. 7-5: Mouth breathing habit may lead to gingivitis (Courtesy Dr Abhishek Soni).

- *Bobby pin opening*—seen in teenage girls who open bobby pin with anterior incisors to place them in the hair. This results in notching of incisors and denudation of labial enamel.
- *Other habits*—nail biting, pencil and lip biting (Fig. 7-6), lead to proclination of upper anterior and retroclination of lower anterior teeth.

- *Bruxism*—it may lead to attrition.
- *Tobacco*—tobacco preparations such as khaini, manipuri tobacco, mawa, mishri, pan, snuff, zarda etc should be ask.
- *Smoking*—smoking habits such as bidi, chutta, cigarette, dhumti, hookah, etc.
- *Drinking habit*—drinking alcohol, charas, ganja, marijuana etc.



Fig. 7-6: Lip biting habit of patient (Courtesy Dr Shetty).

- *Patient's appetite*—whether it is regular or irregular? You should ask the type of diet (vegetarian, mixed, spicy food) to the patient.
- *Soft diet*—soft refined diet adheres tenaciously to the teeth and is not removed because of lack of rough edges leading to more dental caries than coarse diet.
- *Coarse diet*—in persons, having coarse diet, there is more evidence of attrition.
- *Carbohydrate content*—increased carbohydrate contents leads to increased prevalence of dental caries. Phosphate diet prevents dental caries.
- *Vitamin deficiency diet*—diet having deficiency of vitamins may cause enamel hypoplasia. Deficiency of calcium and phosphorus during period of tooth formation results in enamel hypoplasia and defects of dentin. Vitamin D can reduce the caries incidence.
- *Fluoride content*—fluoride content may influence the caries process.
- *Bowel and micturition habit*—Is it regular or irregular?
- *Sleep*—insomnia occurs in cases of primary thyrotoxicosis.
- *Social status*—caries, arteriosclerosis, heart disease, and acute appendicitis are more common in middle and high

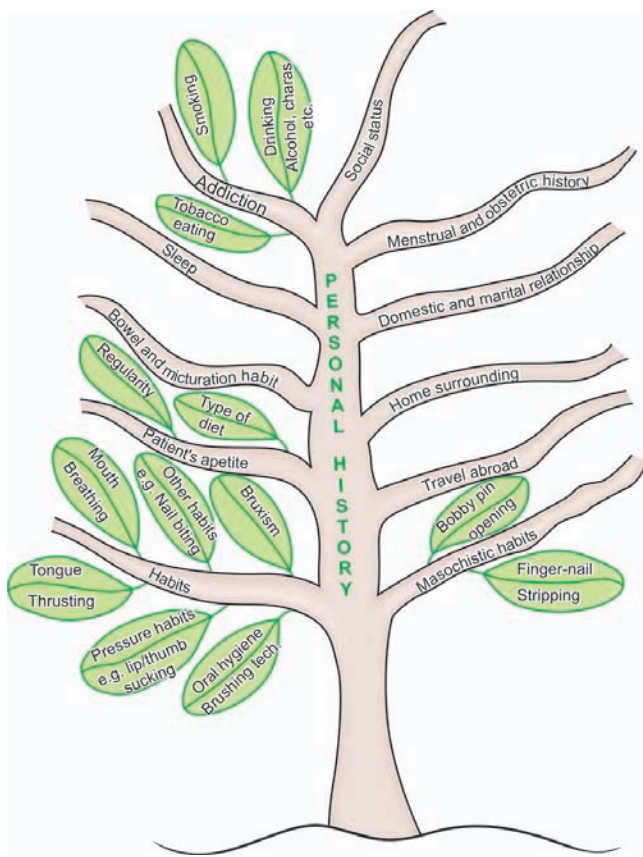


Fig. 7-7: Diagrammatic representation of personal history.

social status individual. Periodontitis, tuberculosis, anemia due to malnourishment is more common in low social status individual.

- **Menstrual and obstetric history**—you should ask about premenstrual tension, presence or absence of pain with periods, whether the patient is taking oral contraceptives, obstetric history, miscarriage, therapeutic abortion. Excessive bleeding may occur during childbirth in cases of hematological disorders. Endocrine diseases may be accompanied by abnormalities of pregnancy, labor and lactation. Cardiac failure may be triggered by circulatory demand of pregnancy and labor.
- **Domestic and marital relationship**—ascertain feelings about other members of family. Interest, hobbies, hopes, fears, the amount of exercise taken. Interest in games and sports. In males, particularly, if unmarried consider the possibility of homosexuality which may be associated with personal and social stresses that may culminate in mental illness.
- **Home surrounding**—whether the surrounding is overcrowded or there is loneliness? What pets are kept?
- **Travel abroad**—whether the patient has lived abroad and if so whether he/she was ill there? Recent travel- e.g., patient may suffer from malaria if he has recently traveled from or even through malaria prone area.

- **Masochistic habit**—some children use the finger nail to strip away the gingival tissue from the labial surface of a lower cuspid. If not discontinued, the marginal and attached gingival tissue will be stripped away exposing the alveolar bone.

Family History

This is very important for many hereditary diseases. Many diseases recur in families like hemophilia, diabetes mellitus, hypertension and heart diseases.

Other Past History

- **Past laboratory investigations**—this will aid in diagnosis of the disease.
- **Previous hospitalizations and operations**—this is important as it can affect the treatment plan of the patient.
- **History of immunization**—you should ask history of immunization regarding chickenpox, mumps and measles. The reason behind this is that, many oral manifestations may mimic above disease.
- **History of allergy**—this is very important. You should ask history of allergy to the drug, specific allergen or any other specific allergy.

Examination

Steps of Clinical Examination

- **Inspection**—it is referred to as passive visualization of the lesion. Intraorally mirror can be used for visualization of structure.
- **Palpation**—this will help for evaluation of structure and to know their consistency, fixity etc.
 - **Bilateral palpation**—this is done to differentiate between symmetrical structures on both side of face (Fig. 7-8).
 - **Bidigital palpation**—in this, two fingers are used to manipulate the tissue. It is used for thinner structure (Fig. 7-9).
 - **Bimanual palpation**—it is done by palms of both hands. In this, one hand is used to support the structure hand and another is used to manipulated the structure. This type of palpation is used in floor of mouth (Fig. 7-10).
- **Auscultation**—it is used to study the movement of TMJ and also used for examination of venous malformation (Fig. 7-11).
- **Probing**—this is used to detect carious lesion.
- **Percussion**—this is used to check whether patient is having periapical infection.



Fig. 7-8: Bilateral palpation is done to palpate the TMJ.



Fig. 7-9: Bidigital palpation is done in case of thinner structure.



Fig. 7-10: Bimanual palpation done in case of floor of mouth.



Fig. 7-11: Auscultation of TMJ is done to examine the movement of TMJ.

General Examination

Build

It can be described as poorly built, moderately built and well built.

- *Asthenic*—it appears as lean and underweight.
- *Sthenic*—they are athletic in appearance.
- *Hypersthenic*—persons have thick muscular and heavy bone structure.
- *Pyknic*—they appear as heavy and rounded.
- *Cachexia*—there is abnormally low tissue mass, occur due to malnutrition.

Pulse

- *Significance*—it is an important index for severity of illness, abnormalities of heart and the vascular system e.g. hypertension and hypotension, shock, fever and thyrotoxicosis.
- *Types of pulse*—pulse can be radial pulse (located on ventral aspect of wrist), branchial pulse (located medial to biceps tendon in the antecubital fossa) and carotid pulse (medial to sternocleidomastoid muscle, inferior and medial to the angle of mandible).
- *Technique*—Assuming that the radial pulse is to be examined, the examiner should take the opportunity to note the warmth of the hand. Find the radial artery a few centimeters proximal to the wrist and just medial to the radius. Palpate the artery wall with the tips of the index and middle fingers. The tips are very sensitive. Avoid the thumb for fear of misinterpreting your own radial pulse. Do not press too hard for fear of obliterating the pulse (Fig. 7-12).



Fig. 7-12: Palpation of radial artery for the examination of pulse of the patient.

- *Points to be noted*—following are particularly noted in pulse:
 - *Rate*—fast or slow (normal 60-100 beats/min).
 - *Rhythm*—regular or irregular, irregularly irregular in arterial fibrillation and regularly irregular in ventricular ectopic.
 - *Volume*—high, low and normal volume indicate pulse pressure (normal pulse pressure is 40-60 mm/Hg). Narrow pulse pressure is in left ventricular failure, obstruction to left ventricle like mitral stenosis, aortic stenosis. Wide pulse pressure in aortic and mitral regurgitation, high output states like pregnancy, anemia etc.
 - *Tension and force*—it indicates diastolic and systolic pressures.
 - *Character*—water hammer pulse of aortic regurgitation, *pulsus paradoxus* of pericardial effusion, *anaerotic pulse* (it is a slow rising twice beating pulse where both waves are felt during systole in carotid artery) in aortic stenosis, *pulsus bisferingus* (it is a rapid rising twice beating pulse where both waves are felt during systole) in idiopathic hypertrophied subaortic stenosis, *dicotic pulse* (it is twice beating pulse where the first percussion wave is felt during systole and second dicotic is felt during diastole) in fever like typhoid, congestive cardiac failure, cardiac tamponade following open heart surgery.
 - *Other factors*—other factors like condition of arterial wall, peripheral pulsations of radial artery, carotid artery, brachial artery should also be seen. Delay of the left temporal pulse compared with right pulse is found in coarctation of aorta.

Blood Pressure

- *Normal blood pressure*—Systolic 120–140 mm Hg and diastolic 80 mm Hg (any above 90 is abnormal). Normal—

120/80 mm Hg. The stroke volume of the heart and stiffness of the arterial vessels control systolic blood pressure. Peripheral resistance controls diastolic blood pressure.

- *Blood pressure variation*—blood pressure varies with emotion, exercise, meal, alcohol, tobacco, bladder distension, temperature and pain.
- *Conditions diagnosed by blood pressure*—conditions diagnosed by blood pressure are hypertension, hypotension and aortic incompetence.
- *Unequal blood pressure*—unequal blood pressure in 2 arms may be due to supravalvular aortic stenosis (right side higher blood pressure), preductal coarctation of aorta (same) and unilateral occlusive disease of aorta.
- *Procedure for blood pressure determination*—blood pressure determination is easily accomplished and is well within the capabilities of every dentist. The procedure is as follows.
 - *Equipment*—the equipment required is stethoscope and sphygmomanometer.
 - *Patient position*—seat the patient and place his right arm on a table or desk so that the forearm is adequately supported at elbow level about the level of heart.
 - *Placement of cuff*—center the rubber balloon of the cuff over the brachial artery (**Fig. 7-13**). The artery is usually palpable about three quarters of the way across the arm going in a lateral to medial direction. Wrap the pressure cuff smoothly and snugly around the upper arm with its lower border 1 to 2 inches above the elbow crease in the antecubital fossa.



Fig. 7-13: Measurement of blood pressure with the help of stethoscope and sphygmomanometer.

- *Pumping of cuff*—palpate the patient's radial pulse and pump the cuff up to above 20 to 30 mm Hg, the point at which the pulse is no longer palpable. It is essential to always record the systolic pressure by palpation to avoid missing the occasional occurrence of silent gap in hypertensive patients.

- *Measurement of systolic pressure*—applies either the bell or the diaphragm of the stethoscope over the artery just distal to the elbow crease and slowly deflates the cuff about 3 mm Hg per heart beat until the first sound appears. Record this first sound as a systolic pressure.
- *Measurement of diastolic pressure*—further lower the pressure in the cuff until the sound disappears. Record this point as the diastolic pressure.
- *Errors*—faulty technique in taking the blood pressure may induce error. If the cuff is applied too loosely and if it is not completely deflated before applying or it is too small for the patient's arm size, the pressure reading obtained will be inaccurately high and will not represent the pressure in the artery at the time of determination. Anxiety and pain are factors that may cause an elevation of the blood pressure of the patient above his true resting level.

Temperature

- *Procedure*—this is normally taken in the mouth or in the axilla by keeping mercury thermometer (**Fig. 7-14**) for a minute. The temperature of mouth is about 1 degree higher than that of axilla. The normal body temperature varies from 36 degree Celsius to 37.5 degree Celsius. The lowest temperature being between 2-4 am and highest in the afternoon. The normal oral temperature is 37°C. Fever or pyrexia is more than 1 degree or any rise above the maximum normal temperature.
- *Conversion of Fahrenheit to Celsius*—temperatures usually taken in Fahrenheit and to convert Fahrenheit into Celsius subtract 32 and multiply by 5/9.



Fig. 7-14: Temperature should be recorded with the help of thermometer.

- *Types of fever*
 - *Continuous fever*—the temperature remains above normal throughout the day and does not fluctuate more than 1 degree in 24 hours. E.g. lobar pneumonia, typhoid and infective endocarditis.
 - *Remittent fever*—The temperature remains above normal throughout the day and fluctuates more than 1 degree in 24 hours e.g. typhoid.
 - *Intermittent fever*—the temperature is present only for some hours in a day and remits to normal for the remaining hours. When the spike occurs daily, it is called as 'quotidian', when every alternate day it is

'tertian', and when every 3rd day it is 'quartian' e.g. malaria, kala azar, pyemia and septicemia.

- *Septic fever*—the temperature variation between peak and nadir is very large and exceeds 5 degrees e.g. in septicemia.
- *Low grade fever*—temperature is present daily especially in the evening for several days but does not exceed 37.8 degree at any time, like in tuberculosis.
- *Hyperpyrexia*—tetanus, malaria, septicemia, heat stroke, encephalitis, and hemorrhage.
- *Benefit of fever*—in some diseases, fever is beneficial. Fever is associated with release of endogenous pyrogen, which activates T cells and thus enhances the host defense mechanism.
- *Harmful effects*—there is hypercatabolism—N₂ wastage and weight loss, fluid and electrolyte imbalance due to sweating, convulsions, brain damage, circulatory overload and arrhythmias.
- *Hypothermia*—hypothermia may be caused by endocrine disorders like hypothyroidism, hypopituitarism hypoglycemia, toxicity like alcohol intoxication, barbiturate poisoning, ketoacidosis, and exposure to cold and autonomic dysfunction.

Respiration

- *Significance*—it is used to assess the condition of the patient under anesthesia and in early postoperative days.
- *Procedure*—you should always check the respiration while checking the pulse of the patient. You should look at the patient abdomen and see how many times he breath. One complete cycle of inspiration and expiration is counted as one. Normal rate—14-18 cycles/minute (**Fig. 7-15**).
- *Tachypnoea* (fast breathing)—it occurs in fever, shock, hypoxia, cerebral disturbances, metabolic acidosis, tetany, and hysteria.



Fig. 7-15: Respiration rate of the patient should be counted while examining the pulse of the patient by looking at the abdomen.

- *Bradypnoea*—slow and deep respiration is seen in cerebral compression.
- *Snoring noise*—paralysis of the soft palate causes an inspiratory snoring noise.
- *Expiratory wheezing*—expiratory wheezing is heard in bronchitis and asthma.

Clubbing

- *Definition*—clubbing is bulbous enlargement of soft part of the terminal phalanges of the nail (**Figs 7-16 and 7-17**).



Fig. 7-16: Lateral view of clubbing showing bulbous enlargement of soft part terminal phalanges on nail (Courtesy Dr Milind Chandurkar).



Fig. 7-17: Clubbing seen in nails of leg of patient (Courtesy Dr Milind Chandurkar).

- *Causes*—it includes:
 - *Pulmonary causes*—like bronchogenic carcinoma, lung abscess, bronchiectasis, and tuberculosis with secondary infection.
 - *Cardiac causes*—like infective endocarditis and cyanotic congenital heart disease.
 - *Alimentary causes*—like ulcerative colitis, Crohn's disease and biliary cirrhosis.

- *Endocrine causes*—like myxedema and acromegaly.
- *Miscellaneous causes*—like hereditary, idiopathic and only in upper limb in heroin addicts due to chronic obstructive phlebitis.
- *Pseudoclubbing*—in hyperthyroidism due to excessive bone resorption there is 'drumstick appearance' of the finger resembling clubbing.

Cyanosis

- *Definition*—cyanosis is the bluish discoloration due to increased amount of reduced Hb (more than 5 mg%) in capillary blood.
- *Types*—it can be central, peripheral, cyanosis due to abnormal pigment or mixed.
- *Sites where peripheral cyanosis is looked for*—nailbed, tip of the nose, skin of the palm and toes.
- *Sites where central cyanosis is looked for*—tongue and other sites mentioned above. Tongue is unaffected in peripheral cyanosis.
- *Causes*
 - *Central*—cardiac causes like congenital cyanotic heart disease and congestive cardiac failure, pulmonary causes like chronic obstructive lung disease (**Fig. 7-18**), collapse and fibrosis of lung, pulmonary obstruction, and high altitude due to low pressure of O₂.
 - *Peripheral*—it includes cold, increased viscosity of blood and shock.
 - *Mixed*—it includes acute left ventricular failure, mitral stenosis and cyanosis due to abnormal pigments like sulfonamides and aniline dye.



Fig. 7-18: Central cyanosis seen in tongue showing bluish discoloration of tongue due to chronic obstructive lung disease.

Icterus

- *Definition*—in jaundice, there is icteric tint of the skin, due to presence of bilirubin, which varies from faint

yellow of viral hepatitis to dark olive greenish yellow color of obstructive jaundice.

- *Site where you should look of Icterus*—the places where one should look for Icterus are sclera of the eyeball (**Fig. 7-19**), nailbed, lobule of ear, tip of the nose and under surface of tongue.
- *Hypercarotenemia*—jaundice may be confused with hypercarotenemia in which yellow pigment of carotene is unequally distributed and is particularly seen in the face, palm and soles but not in the sclera. It occurs mostly in vegetarians.



Fig. 7-19: Icterus seen in eyes of patient who is suffering from jaundice (Courtesy Dr Bhaskar Patle).

Extraoral Examination

Skin

- *Appearance*—changes in appearance, rashes, sores, lumps or itching is looked for and history of sun exposure is questioned.
- *Color*—it is seen for anemia and jaundice.
 - *Generalized pallor*—it is seen in severe anemia. Pallor can be seen in hypopituitarism, shock, syncope, left heart failure and Raynaud's disease.
 - *Lemon yellow tint*—a pale lemon yellow tint in hemolytic jaundice and dark yellow or orange tint in obstructive jaundice is seen.
 - *Yellowness of skin*—yellowness of skin is seen in carotenemia.
- *Texture*—it is thickened, greasy and loose in acromegaly. In dehydration, skin is dry and inelastic so that it can be pinched into ridge. Skin is atrophied with age and sometimes after treatment with glucocorticoids.
- *Signs*—such as petechial hemorrhage indicating blood dyscrasias.
- *Eruptions*—any eruption such as macule, papule, vesicle or bulla.
- *Pigmentation*—pigmentation of Addison's disease affects buccal mucous membrane as well as skin in regions subjected to friction. In Von Recklinghausen's disease, milky coffee pigmentation on the limbs, trunk and face.
- *Edema*—in acute nephritis, edema of face is marked when patient rises in the morning. Dependent edema with pallor and glossy appearance over the swollen part, doughy consistency, and pitting on pressure is characteristic.

Head

- *Headache*—important symptoms like headache should be noted. Following point should be noted in headache.
 - *Nature of headache*—persistent or intermittent and localized or generalized. Unilateral or bilateral or frontal. Associated with unusual disturbances. Familial disorder or induced by stress.
 - *Site*—the exact location of headache.
 - *Radiation*—does it stay in its place or does it move or spread?
 - *Severity*—does it interfere with daily activity or keep you awake at night?
 - *Timing and duration*—when did it start, when does it come and when does it go, has it changed since it began?
 - *Character*—shooting or pricking etc.
 - *Occurrence or aggravation*—what brings it on, and what makes it worse.
 - *Relief*—what makes it better?
- *Head circumference*—hydrocephalus should be suspected when the rate of growth of the head is greater than normal for the sex, age and size of the infant.

Jaws

- *Normal landmark*—normal anatomic landmarks to be identified include mandibular border, angle of mandible, condyle and coronoid process, maxillary bone, lingual notch and maxillary sinus.
- *Tenderness over the jaws*—note any tenderness over the joint or masticatory muscles (temporalis, masseter) while palpating externally over lateral pterygoid and buccinator muscles (distal and lateral to upper molar teeth) and medial pterygoid muscle (pterygomandibular ligament and medial aspect of anterior faucial pillars) with the mouth open.
- *Trauma*—any trauma to head and neck such as injury from motor vehicle or bicycle accident, injury to the side of face and chin should be looked for.

Temporomandibular Joint

The clinical examination for a patient with possible temporomandibular disorder supplements the routine regional head and neck examination. Following point should be noted while examining the temporomandibular joint.

- *History*—medical history about illness, particularly rheumatoid arthritis, degenerative joint disease, osteoarthritis should be asked. Before treatment of temporomandibular joint disorder is initiated, it is necessary to exclude other disorders that might present with similar symptoms like trigeminal neuralgia, multiple sclerosis, glossopharyngeal neuralgia, temporal arteritis, migraine headache, angina pectoris, pulpoperiodontal disease, salivary gland inflammation, duct blockage, otitis, sinusitis and psychogenic pain.
- *Symmetry of face*—the symmetry of the face should be assessed. Gross asymmetry reflects growth disturbances.
- *Interincisal opening*—the maximum interincisal opening of the mouth should be determined, which is, normally, in an adult, 35-50 mm. Measurement also should be recorded for the degree of opening where pain begins.
- *Mandibular movement*—any deviation of the mandible during opening should be noted, along with its severity. The lateral mandibular range of motion is determined by having the patient to occlude the teeth and then slide the jaw in both directions. The range of movement from the midline is measured in mm and any pain along with location and severity is recorded. Normal lateral movement is usually 8-10 mm.
- *Question to be asked to patient*—jaw and joint symptoms should also be discussed with the patient:
 - Does the TMJ click or pop on opening or closing?
 - Has there been limitation in the movement or deviation of the lower jaw on opening?
 - Has the jaw ever locked or dislocated on opening?
 - Has the patient experienced pain and dysfunction in other joints of the body?
- *Palpation of joint*—palpate the joint and listen for clicking and crepitus during the opening and closing of the jaw. Use stethoscope to characterize and locate this sound accurately. Explore the anterior wall of external auditory meatus for tenderness and pain that are usually associated with arthritic changes.
- *Methods of palpation*
 - *Pretragus palpation*—the patient should be requested to slowly open and close the mouth while the doctor bilaterally palpates the pretragus depression with his/her index fingers (**Fig. 7-20**).
 - *Intra-auricular palpation*—it is also performed by inserting the small finger into the ear canal and pressing anteriorly (**Fig. 7-21**).



Fig. 7-20: Examination of TMJ with pretragus palpation method.



Fig. 7-21: Intraarticular palpation method showing finger inserted in the articular area.

- *Significance of palpation*—it is important to perceive, during the pretragus and intra-auricular palpation, whether the condyle moves symmetrically, with the rotation and translation phase being evident. Palpation is also used to detect the tenderness, clicking and crepitus. Subluxation of the joint is also recorded.

Masticatory Muscle

- *Digital palpation*—regional muscles are examined for the tenderness and trigger points using the digital palpation.
- *Masseter muscles*—the masseter muscles are most effectively examined by simultaneously pressing them from inside and outside the mouth in the process of bimanual palpation.
- *Lateral pterygoid muscle*—the lateral pterygoid muscles are evaluated by inserting a finger each behind the maxillary tuberosities, whereas the medial pterygoid is checked by running a finger in an anteroposterior

direction along the medial aspect of the mandible in the floor of mouth.

Lymph Nodes

- **Significance**—normal lymph nodes are difficult to palpate. Nodes draining area of active infection are usually tender; the overlying skin may be warm and red. Gradually enlarging group of nodes in the absence of local infection and inflammation is a significant finding that suggests either systemic disease (infectious mononucleosis, generalized lymphadenopathy associated with HIV infection, lymphoma) and justifies examination for lymphoid enlargement at distant sites such as axilla, inguinal region and spleen to confirm the generalized nature of the process.
- **Inspection of lymph node swelling**
 - **Position**—position is important, as it will not only give an idea as to which group of lymph nodes is affected but also help in diagnosis.
 - **Number**—it is important since in some diseases there is generalized involvement of the lymph nodes like Hodgkin's disease, lymphatic leukemia and lymphosarcoma.
 - **Pressure effect**—swelling and venous engorgement of the face and neck may occur due to pressure effect of the lymph nodes at the root of the neck.
- **Palpation**—the swelling is palpated for number, situation, local temperature, surface, margin and consistency. Enlarged lymph nodes should be carefully palpated with palmar aspect of the fingers. While rolling the finger against the swelling slight pressure is maintained to know the actual consistency.
- **Fixity to underlying tissues**—the lymph nodes of the neck should be examined by standing behind the patient with the patient's neck slightly flexed.



Fig. 7-22: Palpation of submental lymph node is done by standing in front of the patient and patient neck flexed.

- **Palpation of superficial lymph nodes**—palpation of the lymph nodes of the neck is commonly begun with most superior nodes and worked down the clavicle for supraclavicular nodes, anterior to the tragus of ear for preauricular nodes, mastoid and base of skull for posterior auricular and occipital nodes, under the chin for submental nodes (Fig. 7-22) and further posteriorly for submandibular and sublingual lymph nodes.
- **Examination of deep cervical nodes**—to examine the deep cervical nodes ask the patient to sit erect and turn the head to one side to relax the sternomastoid muscle (Fig. 7-23). Use thumb and finger to palpate under the anterior and posterior borders of the relaxed muscle and repeat the procedure on the opposite side. Palpate the posterior cervical nodes in the posterior triangle close to the anterior border of the trapezius muscle. Finally, check for supraclavicular nodes just above the clavicle, lateral to the attachment of the sternomastoid.



Fig. 7-23: Examination of deep cervical lymph nodes is done by relaxing the sternomastoid muscle.

- **Causes of lymph nodes enlargement**
 - **Inflammatory**—acute lymphadenitis, chronic lymphadenitis, septic, tuberculosis, syphilis, filariasis, and lymphogranuloma inguinale.
 - **Neoplastic**—primary (lymphosarcoma), secondary (carcinoma, sarcoma, malignant melanoma).
 - **Hematological**—Hodgkin's disease, non-Hodgkin lymphoma, chronic lymphatic leukemia.
 - **Immunological**—AIDS, serum sickness, drug reaction, SLE and rheumatoid arthritis.

Examination of Other Parts

- **Nose**—severe nasal obstruction leads to breathing through mouth which leads to dryness of mouth which results in persistent sore throat. Infection from nose may spread to the orbit from the adjacent paranasal sinus. Longstanding nasal obstruction suggests a deviated nasal septum. *Epistaxis* (nasal bleeding) may indicate life threatening conditions like cerebral hemorrhage. Nasal discharge may be purulent, bloody, watery or mucoid.
- **Paranasal sinus**—an apical tooth abscess of the upper jaw may drain into the maxillary sinus causing acute sinusitis.
- **Eyes**—visual disturbances, color of the sclera and conjunctivae are looked for anemia and liver diseases. Ocular pain, diplopia and edema of eyelid etc. are also looked for.
- **Ears**—impaired hearing, loss of hearing, discharge from ears, tinnitus.
- **Neck**—check for movement in neck, lump in the neck, cervical lymphadenopathy. If thyroid enlargement is suspected, check if the mass moves up and down with trachea when the patient swallows.
- **External jugular vein**—observe the external jugular vein as it crosses the sternomastoid muscle and with the patient at an angle, approximately 45 degree to the horizontal, note any distension and/or pulsation in the vein. Distension more than 2 cm above the sternal notch is abnormal and is seen in severe right side heart failure.

Intraoral Examination

Intraoral examination is the most important part in dental point of view.

Diagnostic Set of Instrument

Basic diagnostic set which require for intraoral examination are mouth mirror, tweezers, probe, explorer. The use of mirror is mainly for the retraction and indirect vision. Never

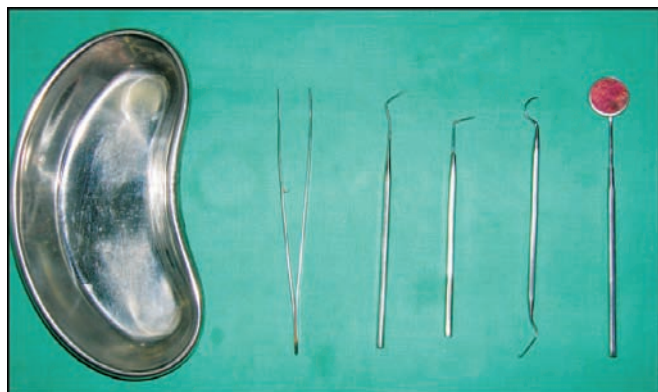


Fig. 7-24: Basic diagnostic set of instrument consist of mouth mirror, probe, tweezers, explorer.

try to directly visualize upper teeth in the patient mouth (Fig. 7-24).

Doctor's Position

Position of doctors is very important while intraoral examination. As there are many professional hazards which occur due to improper position of the doctors.

- **Basic position**—doctor should be either low seated or in standing position. In cases of examining while doctors in standing position, he should stand erect without bending of spinal cord. Patient head should be at elbow level of the doctor. The ideal position is 10 o' clock or 11 o' clock position.
- **Guideline for examination**—you should avoid resting arms over patient shoulder, and maintain comfortable distance from the patient. Eye contact should be maintained with the patient. Drape the patient and avoid keeping instrument on the patient chest.
- **Upper left posterior region**—In this case doctors should stand behind the patient with his left hand going around the patient head (Fig. 7-25).



Fig. 7-25: Doctor's position of upper left posterior region.

- **Upper right posterior region**—doctors should stand at side of patient (Fig. 7-26).
- **Lower right posterior region**—in this case, doctors should stand at side of the patient (Fig. 7-27).
- **Lower left posterior region**—in this case, doctor should stand behind the patient (Fig. 7-28).

Examination of Nondental Structures

Tongue

Inspection

- **The volume of the tongue**—massive tongue due to lymphangioma, hemangioma and neurofibroma.



Fig. 7-26: Doctor's position for upper right posterior region.



Fig. 7-27: Doctor's position for lower right posterior region.



Fig. 7-28: Doctor's position for lower left posterior region.

- *Papillae of tongue*—observe and note the distribution of filiform and fungiform papillae, margins of the tongue, crenation and fasciculation, depapillated areas, fissures, ulcers and keratotic areas.
- *Color*—the white color of leukoplakia, candidiasis (Fig. 7-29) the red color tongue when the leukoplakic plaque gets desquamated, blue color of venous hemangioma. Black hairy tongue due to hyperkeratosis of the mucous membrane in heavy smoker or caused by fungus aspergillus are very characteristic. In jaundice, yellowish tint to the tongue may be seen.



Fig. 7-29: One should note the color of tongue. In this case, color is white due to candidiasis

- *Any crack or fissure*—note the direction of fissure; congenital fissures are mainly transverse whereas syphilitic fissures are usually longitudinal.
- *Swelling and ulcer*—examine the tongue for any swelling or ulcer. If there is a swelling or an ulcer, note the size, shape, extent, margin etc. Also note whether it has extended to the floor of mouth, to the jaw or tonsil. The site of the ulcer is usually characteristic; like the dental ulcer appears on the side of the tongue where it comes in contact with sharp teeth or dentures, tuberculous ulcer on the lip and side of tongue, gummatous ulcer on the dorsum and carcinomatous ulcer occurs usually on the margin of the tongue.
- *Mobility of the tongue*—ask the patient to put the tongue out and move it side ways (Fig. 7-30). Inability to put the tongue out completely is due to ankyloglossia. If the tongue deviates to one side during the protrusion it indicates impairment of nerve supply to that half of the tongue. This is noticed in carcinoma of the tongue, which has damaged the nerve supply of consequent side. If the child with impaired speech fails to protrude the tongue, it is possibly due to tongue-tie; look for short frenum linguae.



Fig. 7-30: Mobility of tongue can be checked by holding it with the hand.

- *Tongue thrusting*—note tongue thrust on swallowing.
- *Central cyanosis*—look for central cyanosis.

Palpation

- *Procedure*—while palpating for induration of the base of an ulcer, tongue should be relaxed and at rest within the mouth. If it is kept protruded, the contracted muscles may give a false impression to induration and lead to error in diagnosis. Induration is an important clinical sign of epithelioma. It may be present in gummatous ulcer but is absent in tubercular ulcer.
- *Points to be noted*—note whether ulcer bleeds during palpation. This usually occurs in malignant ulcer. Palpate carefully for a sharp tooth or tooth placed against an ulcer in the tongue. Palpate the back of the tongue for any ulcer or swelling.

Palate

Inspection

- *Point to look for*—congenital cleft, perforation, ulceration, swelling, fistulae, papillary hyperplasia, tori, recent burns and hyperkeratinization.
- *Cleft examination*—in case of congenital cleft, note the extent of the cleft (involving only the uvula, only the soft palate or part or whole of the hard palate). Whether the nasal septum is hanging free or attached to one side of the cleft (**Fig. 7-31**).
- *Perforation*—perforation of the hard palate is usually caused by gumma.
- *Scar of operation*—any scar of the operation around or history of operation, as sometime a hole may persist after an operation for closure of congenital cleft.

Palpation

- *Point to look for*—tender, fluctuating swelling close to alveolar process is an alveolar abscess. Soft swelling in the midline of the hard palate is gumma.



Fig. 7-31: Note for any cleft in the palate while examining. In this case, cleft is due to cleft palate.

Lip

Inspection

- *Point to be noted*—note lip color, texture and any surface abnormalities, angular or vertical fissures.
- *Cleft*—cleft lip (**Fig. 7-32**), either complete or incomplete, bilateral complete cleft lip in which there is also cleft palate and protuberant pre-maxilla. Cleft between maxilla and side of the nose.
- *Pigmentation*—pigmentation of lip and buccal mucosa in Addison's disease. In Peutz Jeghers syndrome, melanin pigmentation of the lip and oral mucosa occurs.
- *Chancre*—chancre on lip is a painless ulcer with dull red color.
- *Ectopic salivary neoplasm*—ectopic salivary neoplasm is seen in upper lip as slow growing, lobulated tumor.
- *Macrochelia*—thickening of the lip which often involves the upper lip.
- *Cheilitis*—in cheilitis glandularis, lower lip becomes enlarged, firm and everted. In cheilitis granulomatosa, there is diffuse swelling of the lower lip.



Fig. 7-32: Cleft lip should be properly examined in patient.

Palpation

- *Point to be noted*—benign neoplasm is firm and lobulated while carcinoma of the lip is hard in consistency. Chancre is rubbery hard whereas carcinoma is stony hard. Mucous retention cyst is seen on the inner surface of the lip. In cheilitis granulomatosa, the swelling is soft usually exhibiting no pitting on pressure.

Floor of Mouth**Inspection**

- *Procedure*—ask the patient to open his mouth and to keep the tip of tongue upward to touch the palate. This will expose the floor of mouth.
- *Point to be noted*—any swelling, red-white patches in the floor of mouth should be noted.
- *Color of swelling*—when a swelling is present note its color. A ranula appears as a unilateral bluish translucent cyst over which Wharton's duct can be seen. In hemangioma swelling is red in color (**Fig. 7-33**).
- *Site of lesion*—a sublingual dermoid lies exactly in the midline and may extend into the submental region. A deeper plunging ranula may have cervical prolongation into the submandibular region.
- *Ankyloglossia*—in ankyloglossia there is fusion between the tongue and floor of mouth.



Fig. 7-33: Swelling in floor of mouth is due to hemangioma.

Palpation

- *Point to be noted*—Ranula is a fluctuant swelling with positive translucence. Sublingual dermoid which is not translucent but tense and fluctuant swelling is seen on the midline. Carcinoma of the floor of mouth is revealed by its indurated base and probable fixation to the underlying structures.

Cheek**Inspection**

- *Point to be noted*—examine the inside of the cheek for aphthous ulcer, leukoplakia (**Fig. 7-34**), and mucous cyst,

lipoma, mixed salivary tumor, papilloma and carcinoma.

- *Pigmented patches*—pigmented patches are seen in Addison's disease and in Peutz Jegher's syndrome.
- *White lesion*—it can be seen in pronounced linea alba, leukoedema, hyperkeratotic patches.
- *Red lesion*—it can be present in ulcer, nodule, scar, and malignancy.

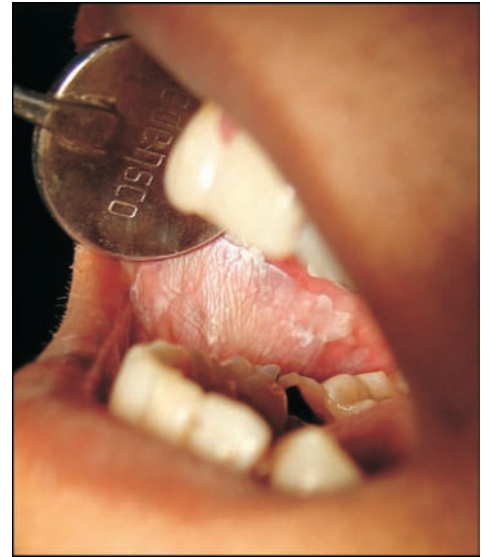


Fig. 7-34: White patch seen on cheek mucosa due to leukoplakia

Palpation

- *Point to be noted*—mucous cysts have smooth surface and are movable over the deeper structures. Fluctuation can be elicited by pressing on the top of the cyst while the sides are palpated by other 2 fingers. Papilloma is a solid tumor with irregular surface and is mobile over the deeper structures. Carcinoma is fixed and indurated. Palpate the muscles of cheek.

Tonsil and Pharynx

- *Point to be noted*—note the color, size and surface abnormalities of the tonsil. Palpate tonsil for discharge and tenderness. Note restriction of oropharyngeal airway. Observe the faucial pillars for nodules, red and white patches (**Fig. 7-35**), lymphoid aggregates and deformities. Observe the postpharyngeal wall for swelling, nodules, lymphoid hyperplasia, hyperplastic adenoid, and postnasal discharge.

Salivary Glands—Parotid Gland**Inspection and palpation**

- *Swelling*—keep in mind the position of the parotid gland, which is below, behind and slightly in front of the lobule of the ear. A swelling of the parotid gland thus obliterates



Fig. 7-35: Redness in tonsillar area should be noted.

the normal hollow just below the lobule of the ear. Position is important since many of the lymph node swellings are mistaken for parotid gland tumor and vice versa. Note the extent, size, shape and consistency. Whether the swelling is fixed to the masseter muscle or not, is confirmed by asking the patient to clench his teeth and mobility is tested over the contracted masseter muscle.

- *Skin over the parotid gland*—in case of parotid abscess, the skin becomes edematous with pitting on pressure. Fluctuation is a very late feature of parotid abscess as there is strong parotid fascia over the parotid gland. The skin is warm and tender. Any scar or fistula should be looked for. If there is malignancy check if there is infiltration of the skin by the tumor.
- *Duct*—Stensen's duct on the buccal surface of the cheek is opposite to the crown of the upper second molar



Fig. 7-36: Stensen's duct opening seen in upper second molar region as a papilla.

(Fig. 7-36). For its proper inspection, retract the cheek with spatula. If there is suppurative parotitis gentle pressure over the gland will cause purulent saliva to come out of the orifice of the duct. Blood will come out in case of malignant growth of the gland. The terminal part of the duct is best palpated bidigitally between the index finger inside the mouth and the thumb over the cheek.

- *Fistula*—if there is parotid fistula, note its position and whether it is in relation to the gland or duct.
- *Examination of facial nerve*—facial nerve is not involved in benign tumor but is involved in malignant growth.
- *Lymph nodes*—preauricular, parotid and submandibular groups of lymph nodes are mostly involved.
- *Movement of the jaw*—it may become restricted if the growth is malignant and has involved the pre-articular tissue of TMJ.

Submandibular Salivary Gland

- *History*—swelling with colicky pain at the time of meals suggests obstruction in submandibular duct. It is tense and painful. Swelling is more often due to lymph node enlargement rather than salivary gland enlargement. If the patient gives the history which is suggestive of a stone in the duct ask the patient to suck a little lemon or lime juice. The swelling will appear at once.
- *Mikulicz's disease*—in Mikulicz's disease; submandibular, parotid as well as lacrimal glands are enlarged.
- *Wharton's duct*—inspection of the Wharton's duct is made by means of torch on the floor of mouth which is situated on either side of lingual frenum. Whether the orifice is inflamed or swollen? If the gland is infected, slight pressure on the gland will exude pus through the respective orifice. If stone is suspected in one duct, saliva will be seen coming out with normal flow from the other orifice while the affected duct orifice remains dry. This is tested by putting dry sweets one on each orifice and some lemon juice is given on the dorsum of the tongue, 2 minutes later the sweets are taken out. The sweet on the orifice of the duct where the stone is suspected will remain dry.
- *Bimanual palpation*—patient is asked to open the mouth; one finger of one hand is placed on the floor of mouth medial to the alveolus and lateral to the tongue and pressed on the floor of mouth as far as possible. The finger of the other hand on the exterior are placed just medial to the inferior margin of the mandible. These fingers are pushed upward; this will help to palpate both the superficial and deep lobes of submandibular salivary gland. This also differentiates an enlarged salivary gland from enlarged submandibular lymph nodes.

Mucobuccal Fold

- *Point to be noted*—observe color, texture, any swelling, and fistulae. Palpate the swelling and tenderness over roots of the teeth and look for tenderness of the buccinator insertion by pressing laterally with finger over roots of the upper molar teeth.

Examination of Dental Structures

Teeth

- *Teeth nomenclature*—various nomenclature systems used are as follows
 - *Zsingingdy's and palmar method*—it is the oldest method of recording teeth. The oral cavity is divided into four quadrants. For deciduous teeth and permanent teeth dental formula is as follows

Deciduous teeth	Permanent teeth
EDCBA ABCDE	87654321 12345678
EDCBA ABCDE	87654321 12345678

- *Universal system*—the primary teeth in the maxillary arch are designated by letter A through J, beginning with the right second molar. The teeth in mandibular arch are designated by letter K through T beginning from the left second molar. Similarly for the permanent teeth numbers 1 to 32 are used.

Permanent teeth—

1 2 3 4 5 6 7 8	9 10 11 12 13 14 15 16
32 31 30 29 28 27 26 25	24 23 22 21 20 19 18 17

Deciduous teeth—

A B C D E	F G H I J
T S R Q P	O N M L K

- *F.D.I (two digit system)*—in it first digit indicates the quadrant as 1 to 4 of the permanent teeth and 5-8 for primary teeth. The second digit indicates the tooth in the quadrant. Thus, deciduous upper right second molar is denoted as 55 and pronounced as five-five and not fifty five and first mandibular left permanent molar is denoted as 36.

Permanent teeth—

18 17 16 15 14 13 12 11	21 22 23 24 25 26 27 28
48 47 46 45 44 43 42 41	31 32 33 34 35 36 37 38

Deciduous teeth—

55 54 53 52 51	61 62 63 64 65
85 84 83 82 81	71 72 73 74 75

- *Dane system*

Permanent teeth—

+8+7+6+5+4+3+2+1	+1+2+3+4+5+6+7+8
-8 -7-6-5 -4-3-2 -1	-1-2 -3-4 -5-6-7-8

Deciduous teeth—

+05+04+03+02+01	+01+02+03+04+05
-05-04 -03-02 -01	-01 -02-03-04-05

- *Point to look for*—you should look for caries (pit and fissure, smooth surface caries, cervical caries), defective restoration or recurrent caries. You should also note missing and supernumerary teeth (Mesiodens, paramolar, distomolar). Presence of root piece of badly carious fractured teeth. You should also look for over-retained deciduous teeth, impacted teeth, ankylosed teeth, fusion of teeth, Talon's cusp, dens evaginatus, taurodontism, and anodontia, enamel hypoplasia, mottled enamel, neonatal teeth, eruption sequestra, delay eruption, attrition, abrasion, erosion, sclerosis, pulp calcification, resorption of teeth, hypercementosis, plaque, calculus and stains.
- *Difference between deciduous and permanent teeth*—it is described in **Table 7-8**.
- *Stains and calculus*—stains and calculus should be recorded in the finding. If calculus is present, there are always chance that gingival inflammation is present as calculus act as local irritant. Different grading for stains and calculus is as follows
 - +—stain and calculus involving only cervical portion of teeth (**Fig. 7-37**).
 - ++—stains and calculus extending upto the middle third of teeth (**Fig. 7-38**).
 - +++—stains and calculus involving whole of the facial or lingual surface (**Fig. 7-39**).



Fig. 7-37: Stains showing involvement of cervical portion only.

- *Percussion test*—this test enables to evaluate the status of the periodontium surrounding a tooth. There are two types of percussion tests carried out i.e. vertical percussion and horizontal percussion test.
 - *Vertical percussion test*—if vertical percussion test is positive, it indicates periapical pathology. To carry



Fig. 7-38: Stains ++ showing involvement upto middle third of the patient. In this patient there is also calculus + on the lingual surface.



Fig. 7-39: Stains +++ and calculus ++ in this patient.

out vertical percussion test the tooth is struck a quick, moderate blow, initially with low intensity by the finger and then with increasing intensity by using the handle of an instrument, to determine whether the tooth is tender. First you have to tap adjacent teeth first and then tap an affected one. The patient response should be taken from patient body movement, reflex reaction of eye. If tooth is tender patient will blink his eyes due to pain (**Fig. 7-40**).

- **Horizontal percussion test**—if horizontal test is positive it indicates periodontium associated problems. In case of horizontal percussion test, the procedure is same but direction of blow is in horizontal direction (**Fig. 7-41**).
- **Mobility and depressibility test**—it is used to evaluate the integrity of the attachment apparatus surrounding the tooth.



Fig. 7-40: Vertical percussion test showing tapping of teeth in vertical direction.



Fig. 7-41: Horizontal blow should be given in horizontal percussion test.

- **Mobility test**—the test consists of moving a tooth laterally in its socket by using the finger or preferably in the handles to two instruments. The objective of this test is to determine whether the tooth is firmly or loosely attached to its alveolus.
- **Depressibility test**—similarly, the test for depressibility consist of moving a tooth vertically in its socket. When depressibility test is positive chances of retaining the tooth range from poor to hopeless.
- **Pathologic mobility**—it results from inflammatory process, parafunctional habits or iatrogenic induced situation.
- **Adaptive mobility**—occurs due to anatomic factors such as short roots and /or poor crown to root ratios that are normally directly contributory to mobility.
- **Measurement of tooth mobility**—there are different ways to measure tooth mobility. They are as follows:

Table 7-8: Difference between primary and permanent teeth

Primary teeth	Permanent teeth
The primary teeth are 20 in number	The permanent teeth are 32 in number
The crowns of primary anterior teeth are wider mesiodistally than the cervicoincisally	The cervicoincisal dimension of the permanent teeth are more as compared to mesiodistal dimension
The crown of the primary teeth have prominent cervical ridge on the buccal surface	The buccal surface of the permanent teeth do not have marked cervical ridge as that of the primary teeth
The roots of the primary teeth are narrow and thin	The roots of permanent teeth are more bulky as compared to that of the primary teeth
The crown and roots of the primary teeth show marked cervical constriction	The permanent teeth do not have such as marked cervical constriction
The roots of the primary molar flare out from the cervical third	The roots of the permanent molars do not possess flare as that of the primary teeth
The furcation is close to the cervical line resulting is a short root trunk	The furcation is at a distance from the cervical line resulting in longer root trunk
The primary teeth are lighter in color because of thinner enamel and different refractive index of the enamel	The permanent teeth are yellowish in color because of thicker and more translucent enamel
Both enamel and dentin thickness is less as compared to the permanent teeth	Both enamel and dentin are thicker as compared to the primary teeth
Pulp chambers are larger in size	Pulp chambers are smaller in size
Pulp horns are at higher level	Pulp horns are at a lower level as compared to the primary teeth
Enamel rods are inclined occlusally in the cervical third	Enamel rods are inclined cervically in the cervical third
Number of accessory canals are more	Accessory canals are less in number
Broader and flatter contact areas	Contact areas are less broad

- According to Glickman
 - Grade I—slightly more than normal.
 - Grade II—moderately more than normal.
 - Grade III—severe mobility faciolingually and/or mesiodistally combined with vertical displacement.
- According to Millar—2nd method was developed by Millar (1950) in which tooth is held firmly between 2 instruments and move back and forth and mobility score is noted (Fig. 7-42) as follows
 - 0—it denotes no detectable movement when force is applied other than what is considered normal (physiologic) motion.
 - 1—it indicates mobility greater than normal.
 - 2—mobility up to 1 mm in buccolingual direction.
 - 3—mobility greater than 1 mm in a buccolingual direction combined with the ability to depress the tooth.
- Dental arch irregularities—it classified by Angle as follows
 - Class I—arch in normal mesiodistal relationship, i.e. mesiobuccal cusp of the maxillary first permanent molar occludes in the buccal groove of mandibular first permanent molar (Fig. 7-43). Dewey's modification of class I Angle classification is as follows.
 - Type I—crowded anterior teeth.
 - Type II—proclinated maxillary incisors.
 - Type III—anterior cross bite (Fig. 7-44).
 - Type IV—posterior cross bite.
 - Type V—the permanent molar has drifted mesially due to early extraction of second deciduous molar or second premolar.
 - Class II—mandibular arch distal to normal in its relation to the maxillary arch, i.e. distobuccal cusp of upper first permanent molar occludes in the buccal groove of lower first permanent molar (Fig. 7-45).
 - Div I—proclination of upper incisors, abnormal muscular activity.
 - Div II—lingually inclined upper central incisors and labially tipped upper lateral incisors overlapping the central incisors.
 - Class II subdivision—Class II molar relationship on one side and class I on other side.
 - Class III—mandibular arch mesial to normal in its relation to the maxillary arch, i.e. mesiobuccal cusp



Fig. 7-42: Mobility of tooth is checked by handles of two instruments.



Fig. 7-43: Class I molar relationship showing mesiobuccal cusp of maxillary permanent molar occlude in buccal groove of mandibular first molar.



Fig. 7-44: Anterior cross bite seen in patient (Courtesy Dr Shetty).



Fig. 7-45: Class II molar relationship showing mandibular arch distal to its normal relationship.

of maxillary first permanent molar occluding in interdental space between mandibular first and second molar (**Fig. 7-46**).

- *Class III subdivision*—class III on one side and class I on other side.



Fig. 7-46: Class III malocclusion showing mandibular arch mesial to normal.

- *Occlusion in deciduous dentition*—it is evaluated by following ways
 - *Flush terminal plane*—the distal surface of maxillary and mandibular second molar are in straight plane.
 - *Mesial step*—distal surface of mandibular second molar is more mesial to maxillary second molar.
 - *Distal step*—distal surface of mandibular second molar is more distal to maxillary second molar.
- *Point to look for in dental arch*—orthodontic anomalies, abnormal jaw relationship. Occlusal interference.
- *Presence of periodontal pocket*—it can be true or false pocket. The probe should be inserted parallel to the vertical axis of the tooth and 'walked' circumferentially around each surface of each tooth to detect the areas of deepest penetration (**Fig. 7-47**).
- *Furcation involvement*—the term furcation involvement refers to the invasion of the bifurcation and trifurcation of multirrooted teeth by the periodontal disease. The extent of involvement is determined by exploration with Naber's probe, along with simultaneous blast of warm air to facilitate visualization. There are different grade of furcation involvement which are as follows:
 - *Grade I*—it is incipient bone loss. Clinically only slight catch occurs in furcation area while probing.
 - *Grade II*—partial bone loss. In this case probe can pass in the furcation area upto middle.
 - *Grade III*—total bone loss with through and through opening of the furcation. Probe can pass through and through.
 - *Grade IV*—it is similar to grade III but with gingival recession exposing the furcation to view (**Fig. 7-48A**).



Fig. 7-47: Probe should be inserted parallel to the vertical axis of tooth in periodontal pocket examination.



Fig. 7-48A: Grade IV furcation involvement which is associated with gingival recession.

- **Pulp testing or vitality testing**—there are four types of pulp testing:
 - **Electric pulp testing**—the electric tester, when testing for pulp vitality uses nerve stimulation. The objective is to stimulate pulpal response by subjecting the tooth to an increasing degree of electric current. A positive response is an indication of pulp vitality and helps in determining the normality or abnormality of that pulp. No response to the electrical stimulus can be an indication of pulp necrosis. The procedure is as follows:
 - **Describe test to patient**—describe the test to the patient in a way that will reduce anxiety and will eliminate bias response.
 - **Isolation of teeth**—isolate the area of teeth to be tested with cotton rolls and saliva ejector and air dry all teeth.

- **Apply the electrolyte**—apply an electrolyte (toothpaste) on the tooth electrode, and place it against the dried enamel to the crown's occlusobuccal or incisobuccal surface. It is important to avoid contacting any restoration on the tooth or the adjacent gingival tissue with the electrolyte or the electrode; this will cause a false and misleading response.
- **Retract the patient's cheek**—retract the patient's cheek away from the tooth electrode with the free hand. This hand contact with the patient's cheek completes the electrical circuit.
- **Turn the rheostat**—turn the rheostat slowly to introduce minimal current into the tooth and then slowly increase it and ask the patient to indicate when sensation occurs by using words as "tingling" or 'warmth'. Record the results according to the numeric scale on the pulp tester (**Fig. 7-48B**).

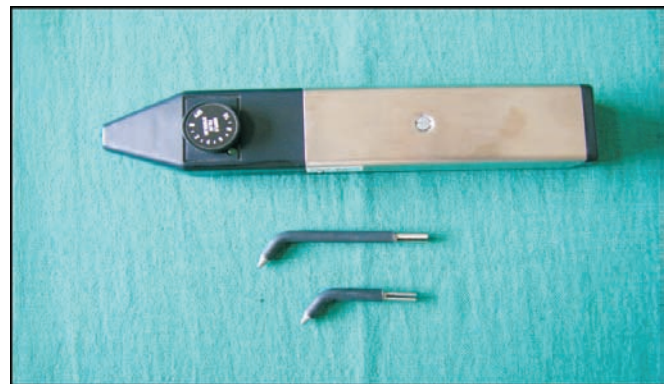


Fig. 7-48B: Electric pulp tester is used for vitality testing of pulp.

- **Thermal testing**—these tests involve the application of cold or heat to a tooth, to determine sensitivity to thermal changes. A response to cold indicates a vital pulp, regardless of whether that pulp is normal or abnormal. An abnormal response to heat usually indicates the presence of a pulpal or periapical disorder requiring endodontic treatment.
- **Heat testing**—the area to be tested is isolated and dried, warm air is directed at the exposed surface of the tooth and the patient's response is noted. If a higher temperature is required then one should use a hot burnisher or hot gutta purcha.
- **Cold testing**—cold air can be directed against the crown of the previously dried tooth and also at the gingival margin. If no reaction occurs the tooth can be isolated under the rubber dam and sprayed with ethyl chloride, which evaporates so rapidly that it absorbs heat and thereby cools the tooth. A more common method is using cotton pellet saturated with

ethyl chloride on the tooth being tested. Another simple method is to wrap a sliver of ice in wet gauze, and to place it against the facial surface of the tooth and to compare to reaction of control tooth.

- **Anesthetic testing**

- *When it is required*—this test is restricted to patient which is in pain at the time of the test, when the usual test has failed to enable one to identify the tooth. The objective is to anesthetize a single tooth at a time until the pain disappears and is localized to specific tooth.
- *Infiltration of teeth*—using either infiltration or the intraligament injection injects the most posterior tooth in the area suspected of being the cause of pain. If pain persists when the tooth has been fully anesthetized, anesthetize the next tooth mesial to it and continue to do so until the pain disappear.
- *Inferior alveolar nerve block*—if the source of the pain cannot be determined, whether in maxillary or mandibular teeth, an inferior alveolar injection should be given.
- *Test cavity*—it is performed when other method of diagnosis have failed. It is made by drilling through the enamel and dentin junction of an un-anesthetized tooth. The drilling should be done at slow speed and without a water coolant. Sensitivity and pain felt by the patient is an indication of pulp vitality. Sedative cement is placed in the cavity and search for the source of infection continues. If no pain is felt, cavity preparation continues until the pulp chamber is reached. If the pulp is completely necrotic, endodontic treatment can be continued in many cases without anesthesia.

Gingiva

- **Color**—normal is coral pink; physiological pigmentation of melanin may be seen.
- **Size**—it corresponds to the sum total of the bulk of cellular and intercellular elements and their vascular supply. Alteration in size is a common feature of gingival disease.
- **Contour**—it depends upon shape of the teeth and their alignment in the arch, location and size of the area of proximal contact and dimension of the facial and lingual gingival embrasures (**Fig. 7-49**).
- **Shape**—it is governed by the contour of proximal tooth surface and the location and shape of gingival embrasures.
- **Consistency**—firm and resilient with exception of free margin.
- **Surface texture**—orange peels appearance or stippled.
- **Position**—the level at which the gingival margin is placed on the tooth.



Fig. 7-49: Irregular shaped contour of gingiva in the patient who has got misalignment of teeth. Gingiva is of normal color.

- **Earliest sign of periodontics**—earliest sign of periodontitis is a deep red line along the free edge of gum. Vincent stomatitis is an inflammatory condition of the gingiva.
- **Cancrum oris**—cancrum oris starts with painful, purple red indurated papule found on the alveolar margin in the region of the molar or premolar teeth which later on ulcerates to rapidly expose the underlying bone and extends to the cheek and lip. Swollen gums, dental abscess and blue line are seen in lead poisoning.
- **Bleeding gums**—as the age advances, there is recession of the gingiva. The gums may bleed, be swollen, spongy and tender in scurvy. Gums may bleed in uremia but may not as spongy as in scurvy. Epulis is the swelling of the alveolar margins of the gums.
- **Gingivitis**—if there is inflammation of the gingiva, it is called as gingivitis. It may be acute or chronic, localized or generalized. Signs of gingivitis are bleeding on slight probing, red, spongy, swollen and edematous gingivae.
- **Periodontitis**—if there is inflammation of the periodontium, that is gingiva, alveolar bone, cementum, and periodontal ligament, it is called as periodontitis. Signs of it include all the sign of gingivitis, gingival recession and alveolar bone loss, foul smell, pocket formation.

Examination of Swelling

Inspection

- **Situation**—a few swellings are peculiar in their position such as dermoid swelling which is mostly seen in the midline of the body or on the line of fusion of the embryonic process. Nasopalatine cyst is common in maxillary anterior region.
- **Color**—the color of swelling should be noted. Color of swelling lead to some clue in the diagnosis.
 - **Black color**—it is of benign nevus and melanoma; red purple color of hemangioma.

- **Pink**—normal color is pink as healthy stratified squamous epithelium is semitransparent and hence, the red color of blood in the extensive capillary bed beneath although somewhat muted shows through. Transitory pink color to whiter appearance is caused due to increased thickness of epithelium layer which occurs due to increased retention of keratin. Masticatory mucosa which is subjected to greatest mechanical stimulation appears light pink, e.g. buccal mucosa, vestibule, floor of mouth and ventral surface of tongue.
- **White**—many pathological variations will impart white color to mucosa. Clinician must be familiar with it.
- **Red**—mucosa changes from pink to red because of thinning of epithelial covering, increased vascularity, and dissolution of part of the collagen content of subepithelial tissue.
- **Yellow**—it is due moderate distribution of adipose tissue contained in connective tissue just beneath the basement membrane e.g. Fordyce's granule.
- **Brownish, bluish or black**—it is induced by melanin, hemosiderin or heavy metal.
- **Contour**—diagnostician must be familiar with normal tissue contour in and around the oral cavity to be able to detect any disorder that might alter the usual configuration of the area (**Fig. 7-50**).



Fig. 7-50: Swelling should be observed for contour, extension, shape.

- **Shape**—the shape of the swelling must be noted, whether it is ovoid, pear or kidney shaped, spherical or irregular. Sometimes, student by mistake utter the term circular to describe the shape of the swelling. A swelling cannot be circular as we do not know about the deeper dimension of the swelling. So it is wiser to say spherical to describe the swelling.

- **Size**—on inspection, we can miss the deeper dimension but we can mention vertical and horizontal extension approximately.
- **Surface**—normal mucosa is smooth and glistening except for area of rugae and attached gingiva which demonstrate stippling and pebbling. Surface of pathological mass may be smooth, ulcerated, papillomatous, eroded, keratinized, necrotic, and bosselated.
 - **Smooth surface**—masses that arise beneath stratified squamous epithelium are *smooth* like fibroma, osteoma, chondroma, hemangioma, intradermal and compound nevus, minor salivary gland tumor, cyst, retention phenomenon, lipoma, schwannoma, neuroblastoma, space abscess and bullous pemphigoid.
 - **Ulcerated surface**—malignant counterparts of above lesions are smooth in early phase but after repeated trauma, they become ulcerated and necrotic.
 - **Cauliflower surface**—it is seen in sq. cell carcinoma, irregular numerous branches on the surface of a papilloma.
 - **Corrugated or papillomatous surface**—masses that originate in the stratified squamous epithelium have 'corrugated' or 'papillomatous' surface like papilloma, verruca vulgaris, seborrheic keratosis, keratoacanthoma, verrucous carcinoma and exophytic growth.
- **Edges**—it may be clearly defined or indistinct, sessile or pedunculated.
- **Skin over swelling**
 - **Red or edematous skin**—it will be red or edematous when the swelling is an inflammatory one.
 - **Tense and glossy skin**—the skin becomes tense, glossy with venous prominence when the swelling is a sarcoma with rapid growth.
 - **Black punctum on skin**—presence of black punctum over a cutaneous swelling indicates sebaceous cyst.
 - **Scar**—presence of scar indicates previous operation, injury or previous suppuration.
 - **Peau d orange skin**—sometimes, the skin over a growth looks like the peel of an orange (peau d'orange), which is due to edematous swelling from blockage of small lymphatics draining the skin.
- **Number**—number of swelling should be noted. Multiple swelling (neuro-fibromatosis), solitary swelling (lipoma, dermoid cyst, etc).
- **Pulsation**—the swellings arising from the arteries are pulsatile like aneurysms and vascular growths such as carotid body tumor. The swellings which lie just superficial to the artery in close relation with it will pulsate. It is called as '*transmitted pulsation*'. The pulsations which originate in the arterial wall will give rise to '*expansible pulsation*'.
- **Movement with respiration**—this is observed if you are examining the swelling in neck and abdominal area. Swellings arising from the upper abdominal viscera move with respiration.

- *Impulse on coughing*—the swellings which are in continuity with abdominal cavity, pleural cavity, spinal canal or cranial cavity, will give rise to impulse on coughing.
- *Movement on deglutition*—few swellings which are fixed to the larynx or trachea move during deglutition like thyroid swelling, thyroglossal cyst, subhyoid bursitis, pre- or paratracheal lymph node enlargement etc.
- *Movement with protrusion of tongue*—there are some swellings which move with the protrusion of tongue like thyroglossal cyst.
- *Pressure effect*—an axillary swelling with edema of the upper limb means the swelling is probably arising from the lymph nodes. Wasting of distal limb indicates the swelling to be a traumatic one. Sometimes a swelling may be seen in the neck with venous engorgement.

Palpation

- *Temperature*—temperature of the swelling is best felt by the dorsal aspect of the hand. First, note systemic temperature. Place finger of the hand on skin in the area of concern and finger of other hand on skin on contralateral part of body. It is increased in inflammation as there is increased metabolic rate and increased vascularity of area. It is increased in superficial aneurysm, arteriovenous shunt and large recent hematoma.
- *Tenderness*—inflammatory swellings are mostly tender whereas neoplastic swellings are non-tender. When the patient complains of pain due to the pressure exerted by the clinician, the swelling is said to be tender.
- *Size, shape and extent*—deeper dimensions of the swelling remain unknown during inspection. The vertical and horizontal dimensions are better clarified by palpation. Tissue surrounding and underlying bases should be carefully palpated to determine the maximum extension of lesion into the surrounding tissue. Whether the mass is poorly defined or moderately defined or well define should be ascertained (**Fig. 7-51**).
- *Surface*—with the palmar surface of the fingers the clinician should palpate the surface of the swelling. It can be *smooth* (cyst), *lobular with smooth bumps* (lipoma), *nodular* (matted lymph nodes) and *irregular and rough* (carcinoma).
- *Edges and border*—Margins are palpated with the help of tip of the finger. Border of firm process in loose connective tissue is readily determined than soft process in loose connective tissue. If the surrounding normal tissue is of same consistency as the pathological tissue, the border cannot be determined.
 - *Benign swellings*—it has smooth margins whereas malignant growth has irregular margins.



Fig. 7-51: Swelling should be palpated for the consistency, margin and extension of swelling.

- *Acute inflammatory swelling*—acute inflammatory swellings have ill-defined or indistinct margins.
- *Malignancies*—malignancies have ill-defined borders that are extremely difficult to palpate as malignant tumors infiltrate adjacent tissue by extending many processes of the tumor into surrounding tissue.
- *Tumor with extension*—tumor with its extension elicits inflammatory reaction in adjacent tissue which results in sequelae of fibrosis which in turn reveal irregular and diffuse areas that are inflamed and result in more tenacious binding of tumor to the adjacent tissue by an ill-defined border, fibrous attachment whose limit is impossible to perceive by manipulation.
- *Consistency*—the consistency or degree of firmness of the lesion in contrast to that of its surrounding tissue will also affect the ease with which the lesion itself or its border may be identified (**Tables 7.9 and 7.10**). Following terms are used to describe consistency:
 - *Soft*—easily compressible tissue such as lipoma or mucocele and cyst.
 - *Cheesy*—indicates a somewhat finer tissue that has granular sensation but no rebound.
 - *Rubbery*—tissue that is firm but can be compressed slightly and rebound to normal contour as soon as pressure is withdrawn.
 - *Firm*—tissue such as fiber tissue that cannot be readily compressed.
 - *Bony hard*
 - *Modifying factors*
 - Thick layer of overlying tissue especially muscle or fibrous tissue.

Table 7-9: Consistency of normal tissue or organ

Soft	Cheesy
• Adipose tissue	• Brain tissue
• Fascia	Rubbery
• Veins	• Skin
• Loose connective tissue	• Relaxed muscle
• Glandular connective tissue	• Glandular tissue with capsule
• Minor salivary gland	• Arteries and arteriole
• Sublingual salivary gland	• Liver
Firm	Bony hard
• Fibrous tissue	• Bone
• Tensed muscle	• Enamel
• Large muscle	• Dentin
• Cartilage	• Cementum
	• Cartilage

Table 7-10: Consistency of pathological mass

Soft
• Cyst
• Warthin's tumor
• Papillary cystic adenoma
• Vascular tumor
• Varicosity's
• Cystic hygroma
• Fatty tumor
• Myxoma
• Plexiform neuroblastoma
• Inflammatory hyperplasia
• Emphysema
• Retention phenomenon (mucocoele and ranula)
Cheesy
• Cyst (sebaceous, dermoid and epidermoid)
• Tubercular node
Rubbery
• Cyst under tension
• Lymphoma
• Myxoma
• Myoblastoma
• Aneurysm
• Pyogenic space infection
• Edentulous tissue
• Early hematoma
Firm
• Infection
• Benign tumor of soft tissue
• Malignancy of soft tissue
• Osteosarcoma
• Chondrosarcoma
• Metastatic carcinoma
• Inflammation and infection of parotid and submaxillary salivary gland
• Inflammation and infection of lymph node
Bony hard
• Osteoma
• Exostosis
• Osteogenic sarcoma
• Chondroma
• Chondrosarcoma

- Soft glandular tissue surrounded by dense connective tissue capsule will be perceived more firm.
- Depth in tissue will alter consistency.
- *Fluctuation*—swelling fluctuates when it contains liquid or gas.

- *Technique*—this test can be carried out by one finger of each hand (**Fig. 7-52**). Sudden pressure is applied on one pole of swelling. This will increase pressure within the cavity of the swelling and will be transmitted equally at right angle to every part of its wall. If another finger is placed on the other side of the swelling the finger will raise passively due to increased pressure within the swelling. This means that swelling is fluctuating. This test is always performed in two planes at right angle to each other. The two fingers should be kept as far as apart as the size of swelling will allow.



Fig. 7-52: Fluctuation test is carried out in patient by using two fingers.

- *Paget's test*—in case of very small swelling which can not accommodate two fingers this test can be performed by simply pressing the swelling at the centre (**Fig. 7-53**). The swelling containing fluid will be softer at the center than its periphery while solid swelling will be firmer at the centre than its periphery. It is called as '*Paget's test*'.
- *Fluid thrill*—in case of swellings containing fluid, a percussion wave is seen to be conducted to its other poles when one pole of it is tapped as done in percussion. In case of big swelling, it can be demonstrated by tapping the swelling on one side with two finger while percussion wave is felt on the other side of the swelling with palmer aspect of the hand (**Fig. 7-54**). In case of small swelling three fingers are placed on the swelling and middle finger is tapped with a finger of the other hand, the percussion wave is felt by other two fingers on each side.



Fig. 7-53: Paget's test is performed in case of small swelling.



Fig. 7-54: Fluid thrill is felt by tapping the swelling.

- *Translucency*—the swelling can transmit light through it. For this it must contain clear fluid like water, serum, lymph or plasma. For this test, darkness is essential. In day time this can be done by using roll of paper which is held on one side of the swelling while a torch light is held on the other side of the swelling. The swelling will transmit the light if it is translucent. The torch light should not be kept on the surface of the swelling, but on one side of the swelling, while roll of paper on the other side so that the whole swelling intervenes between the light and the roll of paper.
- *Impulse on coughing*—on palpation, this test corroborates the finding detected in inspection. The swelling is grasped and the patient is asked to cough. An impulse is felt by the grasping hand. Due to coughing, pressure is increased within the abdominal, pleural, spinal and cranial cavities. This increase in pressure is transmitted to the swelling, where it is felt.
- *Reducibility*—this means that the swelling can be reduced and ultimately disappear as soon as it is pressed upon. This is a feature of hernia, varicocele, meningocele etc.
- *Compressibility*—the swelling can be compressed but would not disappear completely like arterial, capillary or venous hemangioma. The most differentiating features between a compressible swelling and a reducible swelling is that in case of latter the swelling completely disappear as the content are displaced within the said cavities and may not come back until and unless an opposite force such as coughing or gravity is applied, but in the case of the former the content are not actually displaced so the swelling immediately reappears as soon as the pressure is taken off.
- *Pulsatility*—a swelling may be pulsatile if it arises from the wall of an artery or lies close to an artery or if the swelling is a vascular one. Two fingers one from each hand are placed on the swelling as far apart as possible. If the two fingers are raised with each throb of the artery, the swelling is a 'pulsatile' one. When the two fingers are not only raised but also separated with each beat of pulse the pulsation is said to be 'expansile'. When the two fingers are only raised but not separated the pulsation is said to be 'transmitted'.
- *Fixity to the overlying skin*—for this the skin is made to move over the swelling. If it is fixed to the skin, the skin will not move. The skin over the swelling is pinched up in different parts, when the skin is not fixed it can be easily pinched up which may not be possible when the swelling is fixed to the skin. Next attempt is made to move the mass independent of underlying tissue.
 - *Benign lesion*—if it is freely movable then it is benign, possibly encapsulated mass originating in loose subcutaneous tissue or submucosal tissue.
 - *Sebaceous cyst*—sebaceous cyst is freely movable over the underlying tissue but is bound to skin.
 - *Fixed swelling*—swelling may be fixed in fibrosis after previous inflammation episode, infiltrating malignant tumor that originated in skin or mucous membrane and has invaded deep structure, malignancy that originates in deep structure and invade the subcutaneous or submucosal tissue and skin or mucosa and malignancy that originates in loose connective tissue and invade both superficial and deep layer.
 - *Mass in neck*—if fluent mass in neck moves up and down as patient swallows then it must be partly attached to hyoid bone, larynx, trachea, thyroid or parathyroid gland. If it is elevated on protrusive movements of the tongue then it is thyroglossal duct cyst.
- *Relation to the surrounding structures*—The tumor arising from the subcutaneous tissue is free from the overlying

skin and from the underlying contracted muscles. In case of tumor arising from the muscle the swelling can be moved when the muscle is relaxed. Swelling about a bone is absolutely fixed even when the overlying muscle is absolutely relaxed, and cannot be moved apart from the bone.

2 Aspiration

The preoperative aspiration of a fluid-filled mass is worthwhile precautionary procedure, as it should eliminate an unpleasant surprise of opening of an innocuous lesion that may be proved to be a dangerous vascular tumor. Usually it should be carried out before surgery to avoid bacterial infection.

- *Straw colored fluid*—it contains cholesterol crystals in wall that are frequently seen as small shiny particles when syringe is transilluminated. It is seen in some odontogenic and fissural cyst.
- *Thick yellowish white and granular fluid*—it is seen in epidermoid and keratocyst in which lamina is filled with keratin.
- *Yellowish cheesy material*—dermoid cyst contains most of dermal appendage and the aspirate is thickest and fills of yellowish cheesy substance.
- *Sebum*—sebaceous cyst yields sebum which is thick homogenous and yellowish cheesy substance.
- *Amber colored fluid*—dark amber colored fluid of thyroglossal duct cyst.
- *Lymph fluid*—it is colorless with high lipid content, appears cloudy and frothy. It is seen in hygroma and lymphoma.
- *Blue blood*—it is seen in early hematoma, hemangioma and varicosities.
- *Brighter red blood*—aneurysm and arteriovenous fistula.
- *Pus*—aspiration of painful warm fluctuant swelling yields pus.
- *Sulfur granules*—in actinomycosis, it yields pus with few yellow granules in it. These are sulfur granules.
- *Clear viscous fluid*—sticky clear viscous fluid yielded in retention phenomenon.

Other Examination

Other examinations of swelling which are not frequently done in dental office:

- *Percussion*—its role is to find gaseous content within the swelling like resonant note over the hernia or to elicit slight tenderness like Brodie's abscess. Hydatid thrills in case of hydatid cyst.
- *Auscultation*—all pulsatile swellings are auscultated to exclude presence of any bruit or murmur.
- *Measurement*—it is done to find out increase in swelling size at definite intervals but is also to find out if there is any wasting distal to the swelling.

- *Movement of nearby joint*—movement of nearby joint to exclude any impairment.
- *Examination of pressure effect*—nerve may be involved by the pressure of swelling. This will cause wasting, paresis or paralysis of the muscle supplied by the nerve with or without sensory disturbance. The arterial pulse distal to swelling is felt. Sometimes swelling may press on the main artery of the limb and cause weak pulse distally. Swelling may even exert pressure on the adjacent bone by eroding it. It is sometimes seen in aneurysm and dermoid cyst on the skull.

Examination of Ulcer

An ulcer is a break in continuity of the skin and epithelium.

Inspection

- *Size and shape*—the size and shape of the ulcer should be noted. This will yield in diagnosis of the ulcer. The size of an ulcer is important in deciding the time which will be required for healing. A bigger ulcer will definitely take longer time to heal than smaller ulcer. To record exactly the size and shape of an ulcer sterile gauze may be pressed on to the ulcer to get measurement.
 - *Tuberculosis ulcers*—they are generally oval in shape but their coalescence may give an irregular crescentic border.
 - *Syphilitic ulcers*—syphilitic ulcers are similarly circular or semilunar to start with but may unite to form serpiginous ulcer.
 - *Carcinomatous ulcers*—carcinomatous ulcers are irregular in size and shape.
- *Number*—tuberculosis, gummatous, varicose and soft chancre may be more than one in number.
- *Position*—it is very important and often gives clue to the diagnosis. Rodent ulcers are usually confined to the upper part of the face above a line joining the angle of the mouth to the lobule of the ear, occurring frequently near the inner canthus of the eye. Malignant ulcers are more commonly seen on the lips, tongue, breast and penis.
- *Edges*—edges of the ulcer should be properly examined (Fig. 7-55).
 - *Spreading ulcer*—in a spreading ulcer the edges are inflamed and edematous whereas in a healing ulcer the edges, if traced from the red granulation tissue in the center towards periphery, will show blue zone (due to thin growing epithelium) and a white zone (due to fibrosis of the scar).
 - *Undermined edge*—it is mostly seen in tuberculosis. The disease causing the ulcer spreads in and destroys the subcutaneous tissue faster than it destroys the skin. The overhanging skin is thin friable, reddish blue and unhealthy.

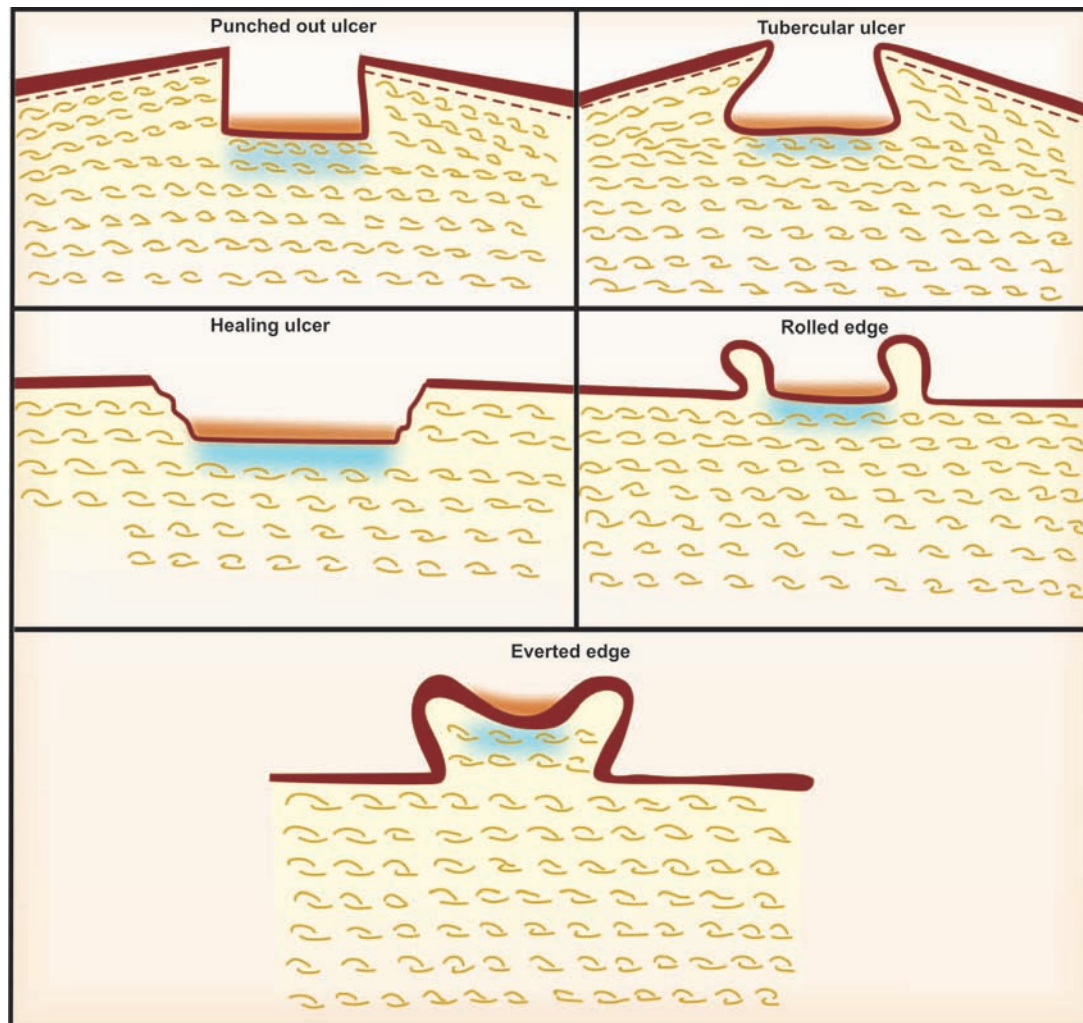


Fig. 7-55: Different types of edges of the ulcer—A diagrammatic representation.

- **Punched out edges**—it is mostly seen in a gummatous ulcer or in a deep trophic ulcer. The edges drop down at right angle to the skin surface as if it has been cut out a punch. It is seen in diseases in which activity is limited to the ulcer itself and does not tend to spread to the surrounding tissues.
- **Sloping edge**—it is seen mostly in healing traumatic or venous ulcers. Every healing ulcer has a sloping edge, which is reddish purple in color and consists of new healthy epithelium.
- **Raised and pearly white beaded edge**—it is a feature of rodent ulcer which develops in invasive cellular diseases and becomes necrotic at the center.
- **Rolled (everted) edge**—it is characteristic feature of squamous cell carcinoma or an ulcerated adenocarcinoma. This ulcer is caused by fast growing cellular disease, the growing portion at the edge of the ulcer heaps up and spills over the normal skin to produce an everted edge (Fig. 7-56).
- **Floor**—this is the exposed surface of the ulcer. One must be very careful to note what is there at the floor of an ulcer. When floor is covered with red granulation tissue, the ulcer seems to be healthy and healing. Pale and smooth granulation tissue indicates a healing ulcer. Wash leather slough on the floor of ulcer is pathognomonic of gummatous ulcer. A black mass at the floor suggests malignant melanoma.
- **Discharge**—the character of the discharge should be noted, its amount and smell. A healing ulcer will show scanty serous discharge, but the spreading and inflamed ulcer will show purulent discharge. Serosanguineous discharge is often seen in a tuberculosis ulcer or a malignant ulcer.
- **Surrounding area**—if the surrounding area of an ulcer is glossy, red and edematous, the ulcer is acutely inflamed. Very often the surrounding skin of varicose ulcer is eczematous and pigmented. A scar or wrinkling in the surrounding skin of an ulcer may well indicate an old case of tuberculosis.



Fig. 7-56: Edges, margins and floor of ulcer should be properly examined. In this case, it has got rolled and everted edges of malignancy.

Palpation

- **Tenderness**—an acutely inflamed ulcer is always exquisitely tender. Chronic ulcers are slightly tender. Neoplastic ulcers are never tender.
- **Edges**—in palpation different types of the edge of the ulcer are corroborated with the finding of the inspection. Marked induration of the edge is characteristic feature of a carcinoma.
- **Base**—the student must understand the difference between the floor (exposed surface within the ulcer) and base (on which ulcer rests and it is better felt than seen). If an attempt is made to pick up the ulcer between thumb and the index finger, the base will be felt. Marked induration of the base is an important feature of squamous cell carcinoma and hunterian chancre.
- **Depth**—you should make assessment regarding depth of the ulcer. It can be recorded in the examination sheet in millimeters.
- **Bleeding**—whether the ulcer bleeds on touch should be checked as it is a common feature of malignant ulcer.
- **Relation with deeper structure**—the ulcer is made to move over the deeper structures to know whether it is fixed to any of these structures. A gummatous ulcer over a subcutaneous tissue or bone is often fixed to it. Malignant ulcer will be fixed to the deeper structure by infiltration.

Examination of Sinus or a Fistula

A sinus is a blind track leading from the surface down to the tissue. Fistula is communicating tract between two epithelial surfaces. Both sinus and fistula are lined by granulation tissue which later may become epithelialized.

Inspection

- **Number**—the majority of fistulae which occur in the body are single. Actinomycosis has multiple sinuses and

sometimes ulcerative colitis may produce multiple fistulae.

- **Position**—diagnosis of sinuses and fistulae can be made only by looking at the position of these sinuses and fistulae.
 - **Preauricular sinus** (due to failure of fusion of the ear tubercles) is situated at the roots of the helix or on the tragus of the pinna, direction of the sinus being upwards and backwards.
 - **Branchial fistula** (due to failure of the fusion of second branchial arch with the fifth) brachial arch is almost always situated at the lower third of the neck in front of sternocleidomastoid muscle.
 - **Osteomyelitis**—a single sinus over an irregular lower jaw is mostly due to osteomyelitis.
- **Opening of sinus**—sprouting granulation tissue at the opening of the sinus suggests presence of foreign body at the depth. The opening of tuberculous sinus is often wide and the margins are thin blue and undermined.
- **Discharge**—in osteomyelitis the discharge is often pus while in actinomycosis discharge is of sulfur granules.
- **Surrounding skin**—there may be scar in the surrounding tissue which may indicate chronic osteomyelitis or previously healed tuberculous sinus. There may be surrounding dermatitis and pigmentation, which are characteristic features of Crohn's disease.

Palpation

- **Tenderness**—is the sinus tender? The sinus from inflammatory source will be tender.
- **Wall of sinus**—it is palpated to note any thickening there. Chronic sinus will have thick wall due to presence of fibrosis surrounding the wall of the sinus.



Fig. 7-57: Gutta percha is used to locate the depth of sinus.

- *Mobility*—is the sinus mobile over the deep structure? Sinuses resulting from osteomyelitis are fixed to the bone, which becomes irregular, thickened and tender.
- *Lump*—presence of lump in the neighborhood of a sinus often indicates tuberculosis lymphadenitis.
- *Probe examination*—this will inform the clinician about the direction and the depth of the sinus. It will also detect presence of any foreign body which will be movable at the depth of the wound and whether fistula is communicating with a hollow viscous or not and whether fresh discharge comes out on withdrawal of the probe or not.
- *Depth of sinus*—it is done with the help of radiograph. A gutta percha point is inserted in the sinus and radiograph is taken (Fig. 7-57). As gutta percha points are radiopaque it will locate the exact direction and depth of the sinus.

Examination of Cranial Nerves

In evaluating patients with oral sensory or motor complaint, it is important to know whether there is any objective evidence of abnormality of cranial nerve function that might relate to the patients oral symptoms. The routine cranial nerve evaluation is systematically carried out according to the sequence of cranial nerve from I to XII.

Olfactory Nerve

- *Procedure*—it is traditionally tested by closing one of the patient nostrils with a finger and asking him if he can smell a strongly scented volatile substance such as coffee or lemon extract. The test is then repeated with other nostril. The patient should sniff strongly to draw the volatile molecules well into the nose (Fig. 7-58).



Fig. 7-58: Smelling of volatile substance from nostril will test for olfactory nerve.

- *Precaution*—such a procedure tests for olfactory nerve function only when the nasal airway is patent to the olfactory receptors and when the substance being tested does not produce a response from the patient solely on the basis of chemical irritation of non-specific somatic sensory receptors in the nasal mucosa.

Optic Nerve

- *Ophthalmoscope*—it is tested by investigation of visual acuity and the visual fields and those trained in the use of the ophthalmoscope can use this instrument to examine the ocular fundus directly for lesions.
- *Wall chart*—visual activity can be tested with the familiar wall chart but can also be evaluated by asking the patient to read print of various sizes in a book or newspaper held at various distance from the patient eye.

Trigeminal Nerve

- *Significance*—it is tested for both motor and sensory functions. A small motor branch of this nerve supplies the muscle of mastication, and strength of these muscles is used as measure of the intactness of their motor supply.
- *Palpation while patient clenches*—the force of contraction and muscle bulk of the masseter and temporal muscles are noted by external palpation of these muscles bilaterally while the patient clenches (Fig. 7-59). Lateral movement of the jaw against the examiner's finger is one test of pterygoid functions.
- *Displacement against resistance*—another useful test of motor power of the masticator muscles is to check their ability to carry out voluntary displacement of the jaw against the imposed resistance of the examiner's hand. Place your thumb on molar table with fingers externally about the body and ramus of mandible. The patient



Fig. 7-59: Test for motor function of trigeminal nerve, patient clenches and doctor palpate masticator muscle.

moves the jaw forward, sideways and upward his head steadied by your other hand.

- *Percussion hammer test*—abnormalities of the jaw jerk may indicate muscular weakness or an abnormality of the proprioceptive reflex arc controlling jaw movement. Press your index finger downward and posteriorly above the mental eminence and lightly strike the finger with a percussion hammer or with one or two finger of the other hand.
- *Graded von Frey hairs*—for testing of trigeminal sensory function ‘graded von Frey hairs’, a series of fine hairs or nylon fibers calibrated according to the force required to bend the filament when it is placed against skin, mucosa or tooth, esthesiometers often designed with a pistol grip to facilitate placement of the points of the instrument on the oral mucosa and calibrated thermal device for application of hot and cold can be used.

Facial Nerve

- *Significance*—the facial nerve is tested for abnormalities of motor function involving the ‘mimetic’ muscle of facial expression and also gustatory disorders. A gustosalivary reflex involves facial nerve gustatory stimuli and increase salivary function.
- *Lemon juice application*—Affected chorda tympani may be associated with failure of salivary flow to increase following application of lemon juice or citric acid to the affected side of the mouth.
- *Motor function test*—motor function of facial nerve is tested by observing facial muscle function in response to request to wrinkle (**Fig. 7-60**) the forehead, frown, close the eyelid tightly, open the mouth, retract the mouth, blow out the cheek, pucker the lips, screw up the nose, whistle and speak.

Glossopharyngeal Nerve

It provides taste fibers to the posterior aspect of the tongue, somatic sensory fibers to the same area of the tongue as well as pharynx and soft palate and motor fibers to the stylopharyngeal muscle that plays only minor role in palatal function, preventing any accurate testing of 9th cranial nerve functions.

Vagus Nerve

- *Significance*—the vagus nerve is the chief motor nerve of the pharynx and larynx and also provides sensory fibers to the pharyngeal and faucial mucous membrane
- *Pharyngeal movement observation*—routine testing is carried out by observation of pharyngeal movements’ i.e. symmetrical elevation of the soft palate and shortening of the uvula when the patient says ‘ah’ and



Fig. 7-60: Patient is asked to wrinkle his forehead for motor function test of facial nerve.

pharyngeal and gag reflex, i.e. contraction of the palate and faucial muscles in response to touching the mucous membrane of the posterior pharynx.

Hypoglossal Nerve

It provides motor supply to the tongue; hypoglossal paralysis causes deviation of the tongue when patient extrudes it. Atrophy of tongue musculature may be noted on oral examination and its muscular tonus by the force with which the patient can push the tongue against either cheek or by evaluation of the tongue jerk.

Establishing the Diagnosis

Provisional Diagnosis

All the records and clinical findings clubbed together, clinician should be able to frame a provisional diagnosis. Clinician should keep in mind the differential diagnosis of the lesion. In provisional diagnosis you should list all those items that either indicate an abnormality or suggest the possibility of significant health problems requiring further evaluation.

Investigations

Laboratory investigations help to come to the final diagnosis, e.g. in case of caries (proximal) the provisional diagnosis will be mesial or distal caries. Radiograph will confirm the diagnosis and help us to differentiate it into incipient, moderate, advanced and severe. To confirm the

Table 7-11: Case history format

Personal information	• Clubbing • Cyanosis	Cheek
Name		• Color
Age / sex	Extra oral examination	• Any ulcer
Case No	• Facial symmetry	Lips
Occupation	• Lip seal	• Color
Religion	• TMJ movements	• Vermillion border
Address	• Lymph node	• Commissures
Date	• Ear	• Ulcer and lesion
Chief complaint :	• Nose	Hard and soft palate
History of present illness	Intraoral examination	• Discoloration
History of past illness	Hard tissue examination	• Papillary hyperplasia
Past medical history	• Teeth present	• Swelling
Past dental history	• Dental arch irregularities	• Ulcer
Personal history :	• Caries	• Any recent burns
• Marital status	• Fractured tooth	• Any lesion
• Educational status	• Tenderness	Floor of mouth
• Oral hygiene habits	• Wasting disease	Gingiva
• Other habits	• Discolored tooth	• Color
Family history :	• Calculus and stains	• Contour
General examination :	• Occlusal facets	• Consistency
• Patients attitude	• Mobility	• Surface texture
• Built	• Furcation involvement	• Position
• Gait	Soft tissue examination	• Size
• Vital signs :	Tongue	• Bleeding on probing
• Temperature	• Size	• Exudate/suppurative
• Pulse	• Dorsum	• Frenal attachment
• Respiratory rate	• Ventral surface	Provisional diagnosis
• Blood pressure	• Margin	Investigation
• Pallor	• Frenal attachment	Final diagnosis
• Icterus	• Distribution of papilla	Treatment planning :
		• Medical / emergency phase
		• Planned treatment

diagnosis, one should proceed with available methods of investigation.

The common methods are radiological, hematological investigations, urine analysis, biochemical investigations, histopathological investigations, microbiological investigations and special investigations like sialography, MRI etc.

Final Diagnosis

The final diagnosis can usually be reached following chronologic organization and critical evaluation of the information obtained from the patient history, physical examination and the result of radiological and laboratory examination.

The final diagnosis usually identifies the diagnosis for the patient's primary complaint first, with subsidiary diagnosis of concurrent problems. Previously diagnosed conditions that remain as actual or potential problems are also included, with comment by 'history' 'previously diagnosed'.

In case of no definite diagnosis is made diagnosis should be written as idiopathic, unexplained, functional and symptomatic.

The patient should be informed of the diagnosis, results of tests and examination.

Making a Treatment Plan and Medical Risk Assessment

The formulation of an appropriate treatment plan will depend on both knowledgeable, experienced, competent clinician and nature and extent of treatment facilities available. Details of medical or surgical treatment, which the patient has received, should be known. Evaluation of any special risks posed by the patient's compromised medical status in the circumstance of the planned anesthetic diagnostic or surgical procedure.

Medical risk assessment is also needed to identify the need of medical consultation and to recognize significant deviation from normal health status that may affect dental management.

American Society of Anesthesiologists Physical Status Classification

- P1—a normal healthy patient
- P2—a patient with mild disease
- P3—a patient with severe systemic disease that limits activity but no incapacitating
- P4—a patient with an incapacitating systemic disease that is a constant threat to life
- P5—a Mir bund patient who is not expected to survive without operation
- P6—a declared brain dead patient whose organs are being removed for donor purpose

Adapted from American Society of Anesthesiologists

Table 7-12: Medical questionnaire to be asked to the patient in medical examination

System	Question to be asked
General	• Are you now or recently been under a physicians care? if so, what is the condition being treated? • Have you been hospitalized during past 2 years? • Do you take prescription or non-prescription medicines on a permanent or non-permanent basis? • Do you have any allergies, if so to what? • Aspirin • Sulphur containing drugs • Penicillin or any other antibiotics • Codeine or any narcotic • Dental anesthetic or • Any specific drug • Does dental treatment make you nervous?
Cardiovascular system	• Chest discomfort on exertion, when eating or at rest • Tightness of the chest • Palpitation • Fainting • Ankle edema • Shortness of breath • High or low blood pressure • Heart murmur.
Respiratory system	• Dyspnea on exertion • Wheezing, coughing, excessive sputum production • Coughing up blood.
Blood disorders	• Prolonged bleeding during cut, tooth extraction or other injury • Do you bruise easily • Required a blood transfusion • Have frequent infections • Have ever been told that you were anemic.
Nervous system	• Head-injury or concussion • Seizures/black-outs/fits • Frequent headaches • Experienced any pain, numbness, or tingling in your face , arms or legs • Had any paralysis.
Metabolic-endocrine system	• Any recent loss or gain of weight • Hyperthyroidism or hypothyroidism • Do your hands sweat excessively • Are you easily fatigued?
Gastrointestinal, hepatic, biliary tracts	• Difficulty in swallowing • Heart-burn or belching • Episodes of nausea and vomiting • Vomited blood • Frequent stomach or abdominal pain • Recently had a change in bowel habits • Frequent episodes of diarrhea • Ever noticed bright-red, or black colored stools.
Genitourinary system	• Urinate frequently • Difficulty or pain during urination • Ever passed brown or red urine • (for females) had abnormal or irregular menstrual period

- Drugs given to the patient, dose and duration of treatment—the drug can be prescribed in following formats.
 - *OD—Omni Die*
 - *B.I.D.—Bis in Die*
 - *B.D.S.—Bis in die sumendus*
 - *T.I.D.—Ter in Die*
 - *T.D.S.—Ter die sumendum*
 - *Q.I.D.—Quater in die*
 - *Q.D.S.—Quater die sumendum*
 - *S.O.S—*as and when required (si. opus sit)
 - *H.S—*Hora somni
 - *Rx—*Take though
- *Prescription writing*
 - *Superscription*—general background information regarding the dentist (name, address and telephone number), the patient (name, address, age) and the date the prescription is written.

- *Inscription*—specific information regarding the drug (generic or proprietary name or both) and the dosage.
- *Subscription*—direction to the pharmacist for filling the inscription (number of capsules or tablets to be dispensed or the volume of liquid), the number of refills allowed and time constraints, directions to be listed on the container label.
- *Transcription*—instruction to the patient, to be listed on the container label.
- *Signature and educational degree of prescribing doctor*—a signature is required by law only for certain controlled substance (schedule II drug).
- *Guidelines for prescription writing*
 - Obtain an accurate and complete patient history, including whether the patient is taking any drugs prescribed by other doctors or any over the counter drugs; both can affect the dosage or effects of the drug being prescribed.
 - Use separate prescription blank for each drug ordered. Avoid using prescription blank with trade names printed on them.
 - Never presign prescription blank and always store blank prescription pads in a secure place.
 - Write out number than using digits so that prescription cannot be altered.
 - Prescribe sufficient drug and at adequate dosing intervals to maintain therapeutic blood level.
 - Keep the record of all drugs prescribed for each patient.
 - Instruction listed on the drug container for the patient must be specific. Full disclosure is verbally given regarding the prescription before the patient leave

the office. Stress the need to consume all of the prescribed medication, as in taking the antibiotics even if the patient is feeling better before the prescribed amount is taken.

- Instruction regarding anticipated side effect as well as the use of alcohol with taking the prescribed drugs should be explained to the patient verbally. Advise the patient to call the office when side effects develop.
- Cost factor should be considered when prescribing medication.
- *In surgical treatment*
 - Types of anesthesia; local or general anesthesia
 - Anesthetics used
 - Name of the anesthetist
 - Name of the surgeon
 - Closure
 - Drainage given or not
- *Follow up*—these resume when the patient is discharged from the hospital and extends till he starts his normal active life.

Suggested Reading

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3. Greenberg, Glick, Ship. Burket's Oral Medicine: BC Decker Inc: 2008.
4. Kay E, Nuttal N. Clinical decision making—an art or a science. Br Dental J 1995;190-3.
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Investigation in Dentistry

Introduction

There are various important investigations which are required for the diagnosis and treatment plan of various disorders related to the oral cavity. Laboratory studies are an extension of physical examination in which tissue, blood, urine or other specimens are obtained from patients and subjected to histological, bio-examination, microbiological or immunological examination. Information obtained from these investigations help in identifying the nature of the disease.

Diagnostic Test for Cancer Detection

The earliest is the diagnosis of the oral cancer, best is the prognosis of the cancer. In cancer, cellular and tissue alteration occurs. This fact is kept in mind for the entire diagnostics test for cancer detection.

Classification

- *Clinical method*
 - Vital staining—toluidine blue method and Lugol's iodine method
 - Vizilite
 - Acridine binding method
- *Photodiagnosis*
 - 5-Aminolevulinic acid mediated fluorescence endoscopic imaging
 - 5-Aminolevulinic acid mediated digitized fluorescence endoscopic imaging
 - Autofluorescence spectroscopy
 - Fluorescence photography
- *Histopathological methods*
 - Biopsy
 - Exfoliative cytology
 - Oral CDx system
- *Molecular methods*
 - Quantification of nuclear DNA content
 - Tumors markers
 - Microsatellite markers.

Clinical Method

Toluidine Blue Staining

The use of Toluidine blue (tolonium chloride) dye as a mouthwash or topical application is currently receiving much attention as an aid to the diagnosis of oral cancer and potentially malignant lesions. The method has good sensitivity with a very low false negative rate. The dorsum of the tongue always stains positively, due to the retention of dye in crevice between the papillae. It is effective in demonstrating dysplasia and early malignant lesion which is not clinically recognizable.

Mechanism

- *Binding with DNA*—it is an acidophilic, metachromatic nuclear dye of thiazine group that selectively stains acidic tissue components particularly nucleic acid such as DNA and RNA. Dysplastic and anaplastic areas which contains more DNA than normal cells.
- *Intracellular canal*—malignant epithelium contains intracellular canals that are wider than normal epithelium, which also facilitates the penetration of dye.
- *Effect on normal epithelium*—most of the epithelial surfaces stain blue after the application of 1% toluidine blue solution but the stain is lost after application of 1% acetic acid solution to normal epithelial surface or benign erythematous lesions on oral mucosa.
- *Effect on benign ulceration*—benign ulceration has well defined uptake of dye at the margins whereas, diffuse marginal pattern is characteristic of dysplasia or malignancy.

Availability

It is readily available to use kit as 3 component system. First contain 1% toluidine blue 10 ml solution. Second and third kit contains 1% acetic acid as pre- and post-rinse solution.

Contents

- Toluidine blue—1 gm
- Acetic acid—10 cc
- Absolute alcohol—4.19 cc
- Distilled water—86 cc
- pH adjusted to 4.5

Technique

- *Rinsing*—initially you ask the patient to rinse the mouth twice with water (20 seconds each). After rinsing with water, ask patient to rinse with 1% acetic acid (20 seconds).
- *Drying of area*—you should gently dry suspicious mucosal areas with gauze. You have to take care not to abrade the tissue while drying.
- *Application of toluidine blue solution*—apply 1% toluidine blue solution to the lesion with a cotton swab.
- *Rinsing*—ask patient to rinse again with acetic acid (approximately 150 ml for one minute). After rinsing with acetic acid, then ask patient to rinse with water.
- *Positive staining*—if the mucosa is stained positive (**Fig. 8-1**), you have to repeat the procedure in one to two weeks. Biopsy of all sites is advised, which stain positive on two successive visits.



Fig. 8-1: Positive toluidine blue staining showing uptake toluidine blue by dysplastic mucosa (Courtesy Zila Pharmaceutical).

Lugol's Iodine Test

It is retained in normal squamous epithelial cells but not in dysplastic or malignant cells of the squamous epithelium. The use of Toluidine blue application in combination with Lugol's iodine solution may offer potential advantage. The Toluidine blue will stain the abnormal (reactive and dysplastic) epithelium whereas, Lugol's iodine solution will bind to glycogen present in normal epithelium.

In the oral tissues, Lugol's iodine has less sensitivity in identifying oral premalignant and dysplastic diseases. But, is of greater specificity.

Contents

- Iodine—2 gm
- Potassium iodide—4 gm
- Distilled water—100 cc

Mechanisms

- *Reaction with glycogen*—Lugol's iodine solution produces a brown black stain by reaction of iodine with glycogen. Iodine is removed by fixation in alcohol and formaldehyde.
- *Effect on proliferating epithelium*—glycogen content is inversely related to the degree of keratosis, suggesting a role of glycogen in keratinization. So when proliferating epithelium is present it is usually poorly stained or unstained.
- *Effect on inflammatory tissue*—there is relationship between the degree of inflammation and glycogen content. So in this case, tissue will stain dark brown.

Vizilite

Vizilite is non-toxic chemiluminescent light that is shined in the mouth. Tissue which are not normal, they glow differently, as compared to normal tissue. This will make them more visible. Vizilite will be more useful for the detection of biopsy proven squamous cell dysplasia which is not seen with naked eye.

Contents

- Vizilite rinse—1% acetic acid solution
- Vizilite capsule—chemiluminescent light stick
- Vizilite retractor—sheath and handle

Mechanism

- *Physics*—normal tissue absorb Vizilite and as a reason they appear dark. This is not the case with dysplastic cells. In case of dysplastic cells nucleus become larger, resulting in reflection of light and gives white appearance.

Technique

- *Application of Vizilite rinse*—acetic acid solution should be applied first. This will result in change in refractile properties of lesion like leukoplakia. The change will occur in the cells as acetic acid is cytoplasmic dehydration agent and there is increased nuclear-cytoplasmic ratio in atypical keratinized mucosa.
- *Visualizing the mucosa with Vizilite*—then chemiluminescent capsule is projected on the mucosa. The abnormal mucosa like dysplastic mucosa will show more reflection of light and will appear whiter as compared to normal mucosa (Fig. 8-2).



Fig. 8-2: In Vizilite testing, dysplastic mucosa will show appearing whiter as compared to adjacent mucosa (Courtesy Zila pharmaceutical).

Acridine Binding Method

In this method, the uptake of acriflavine by desquamated buccal cells is measured. Since the DNA content of the dysplastic cells are more, they will stain more intensely than normal cells.

Photodiagnosis

This optic method is used for the early diagnosis. It is based on fluorescence measurement. Light induced fluorescence (LIF) spectroscopy is useful for the early cancer detection in oral cavity. The principle is that there is endogenous tissue fluorescence of photosensitizer which is accumulated in tumor tissue. Photosensitizer which can be used are m-THPC and delta aminolevulinic acid.

5-aminolevulinic Acid (ALA) Mediated Fluorescence Endoscopic Imaging

- *Source*—aminolevulinic acid is precursor in biosynthesis of heme and it induces the production of endogenous photosensitizer protoporphyrin.
- *Mechanism*—after oral administration of ALA, synthesis of protoporphyrin occurs in dysplastic cells. This will result in fluorescence and can be easily detected.

5-aminolevulinic Acid Mediated Digitized Fluorescence Endoscopic Imaging

It is same method as above with digitalization facility. It has capability of high quality images and it can measure the fluorescence effect of diseased oral tissue.

Autofluorescence Spectroscopy

- *Use*—autofluorescence spectroscopy is non-invasive method. It is used for detection for alteration in the structural and chemical composition of cells. It is also useful for optimal location of biopsy.
- *Mechanism*—autofluorescence occurs due to endogenous fluorophores. It consists of fluorophores from tissue matrix molecule, intercellular molecules like collagen, elastin and nicotinamide adenine dinucleotide phosphate.

Fluorescence Photography

The basic principle behind this is that, by repeated fluorescence photography, it will show reduction and diminution of positive fluorescence associated with cancer regression and vice versa. This is useful as an adjunct aid in the diagnosis of squamous cell carcinoma.

Histopathological Examination

Biopsy

It is a process of surgically removing tissue from a patient for histopathological examination. It is the removal of sample tissue from living individuals. It provides valuable information in determining the prognosis and type of treatment required.

Indications

- *Undiagnosed clinical condition*—after careful clinical examination, if any alteration from normal is seen and it is not possible to identify the condition clinically, a histopathological investigation is necessary.

- *Nature of lesion*—to evaluate the exact histological nature of any soft tissue or intra-osseous lesion.
- *Screening test*—to screen abnormal tissues removed from oral cavity including granuloma and cyst.
- *Detection of malignancy*—to confirm the existence and nature of directly apparent malignancy so that the treatment can be undertaken immediately.
- *Diagnostic test*—diagnostic tests for evaluation of non-neoplastic lesions such as mucosal nodules, papilloma, erosive lichen planus, erythema multiforme, lupus erythematosus, pemphigus, pemphigoid and desquamative gingivitis.

Avoidance of Delay for Biopsy

Biopsy should not be delayed when following feature are present.

- *Rapid growth*—rapid increase in size of the lesion that cannot be explained by inflammation, edema and opening of new vascular channels.
- *Absent local factors*—absence of any recognized irritant, particularly when the lesion is chronically ulcerated or bleeds spontaneously.
- *Fixed lymph node enlargement*—presence of firm regional lymph nodes, especially when they are seen to be fixed to surrounding tissues.
- *Root resorption with loosening*—destruction of tooth roots and loosening of teeth with evidence of rapid expansion of the jaw.
- *History of malignancy*—history of cancer elsewhere in the body, previous history of oral cancer and radiation therapy.

Uses

- *Diagnosis*—biopsy is useful for the diagnosis of pathologic lesions. It also helps in determining neoplastic and non-neoplastic lesions of oral cavity.
- *Grading of tumors*—it aid in determining the grading of tumor.
- *Metastatic lesion*—it is also useful for the diagnosis of metastatic lesions.
- *Recurrence*—for the evaluation of recurrence.
- *Management assessment*—it is useful for the therapeutic assessment of the lesion by differentiating between benign and malignant lesion.

Types of Biopsy

More commonly used biopsies are excisional, incisional, fine needle, punch, scrape and trephine. Some biopsy like bite, cone, core, endoscopic, irrigation, pressure, shave and sponge are less commonly used.

How to Submit Biopsy Specimen

- *Information written on specimen*—the submission of specimen should be accompanied by the date of biopsy, name, age and sex of the patient, the area from where biopsy specimen is taken and brief description of clinical appearance of lesion and associated symptoms, along with tentative clinical diagnosis. The biopsy specimen should not only include the lesion but also the adjacent clinically normal tissue.
- *Sectioning of biopsy specimen*—as soon as possible after the surgical procedure, carefully section the biopsy specimen with a fresh scalpel or razor blades into pieces. Pieces should not be larger than 0.5 cm in diameter, identifying each fragment in relation to the overall specimen and lesion.
- *Avoid surface with iodine*—iodine containing antiseptics should be avoided with surface since they have a tendency to stain certain tissue cells permanently.
- *Fixing solution*—the portion of the biopsy specimen to be used for routine histological study should be placed at once in a suitable fixing solution. Fixative which is used is Karnovsky's fixative (4% paraformaldehyde) and 5% glutaraldehyde in 0.1 M solution for 2 to 4 hours and delivers promptly to the laboratory.
- *Stains*—many different types of stains are used in diagnostic pathology (Fig. 8-3).



Fig. 8-3: Different types of stains used in diagnostic pathology.

- *Submission of teeth*—for the histologic examination of teeth, the apex of tooth should be clipped with a pair of pliers or a small hole should be drilled into the radicular pulp with dental burr to allow penetration of the fixative.
- *Preservation*—the excellent preservation of cellular detail required is obtained by following methods.
- Cutting the specimen into tiny blocks before fixation.
- Use of special fixatives that preserve cellular detail with minimum disruption from rapid dehydration or osmotic shock.

- Post-fixation and processing of tissues in the laboratory after the initial period of pre-fixation (**Fig. 8-4**).

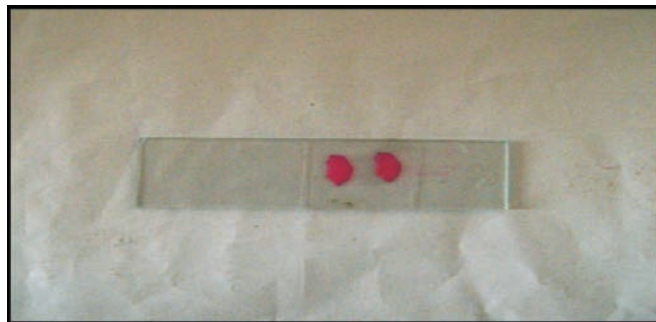


Fig. 8-4: Biopsy slide is prepared and stored properly.

Instrument and Materials

- *Excisional and incisional*—Local anesthesia with vasoconstrictor, scalpel holder and blade. Pointed surgical scissors, tissue forceps and surgical hemostat. Sterile sponge, curved needles and suture, needle holder. Wide mouthed bottle containing 10% formalin.
- *Intraosseous biopsy*—periosteal elevator, bone bur, curette.
- *Aspiration biopsy*—large syringe (10–20 cc) with a large needle.

Excisional Biopsy

Total excision of a small lesion for microscopic examination is called as '*excisional biopsy*'. It is a therapeutic as well as a diagnostic procedure. Normal tissue on the margins of the lesion should be included.

Indications

- *Small lesion*—it is the preferred treatment if, the size of lesion is such that it may be removed along with the margins of normal tissue and wound can be closed primarily. It is usually done in case of lesion smaller than 1 cm.
- *Sessile lesion*—when the lesion is sessile or pedunculated, excisional biopsy can be done.
- *Movable tissue*—tissues which are freely movable and located above the mucosa or just beneath the surface.

Procedure

- *Local anesthesia*—anesthetize the lesion with 2% local anesthetic containing vasoconstrictor. Care is taken not to inject directly into the lesion that is to be removed.
- *Elliptical incision*—with the scalpel, make an elliptical incision on either side of the base of the lesion so that incision line intersected. The blade should be at an angle of 45° towards the center of the lesion.
- *Pulling of tissue*—outward tension is placed on the lesion by means of suture or with the help of tissue forceps

attached at the edge of specimen. Care must be taken not to crush the specimen.

- *Dissection of tissue*—the specimen is now gently dissected out with either a scalpel or a pair of surgical scissors.
- *Keeping in formalin solution*—the tissue must be immediately submerged in 10% formalin solution.
- *Suturing*—surgical site is closed with either silk or absorbable sutures placed approximately 5 mm apart.

Incisional Biopsy

Incisional biopsy can be performed by removing a wedge shaped specimen of the pathological tissue along with surrounding normal zone.

Indications

- *Large lesion*—if the lesion is large and diffuse and extends deeply into the surrounding tissue so that total removal cannot be obtained easily with local anesthesia, an incisional biopsy is indicated.
- *To determine nature of treatment*—lesions in which diagnosis will determine whether the treatment should be conservative or radical.

Procedure

- *Selection of site*—the selection of site is important. It is best to select the site away from an obvious ulceration or area of necrosis, as these are areas of intense inflammation which make interpretation difficult.
- *Local anesthesia*—the tissue around the specimen is infiltrate with 2% local anesthetic.
- *Elliptical incision*—with a scalpel, make an elliptical incision encompassing the selected area of the lesion. The incisional lines must be deep enough to include underlying connective tissue to the level of muscle or bone.
- *Suturing*—suture is inserted through the end; upward tension is applied while tissue sample is dissected out.

Intraosseous Biopsy

It is less frequently performed. It may be in the form of exploratory curettage in which the representative tissue is obtained to determine the nature of large radiological alterations.

Procedure

- *Selection of site*—after correlating the radiographs with overlying anatomical structures, the site of which the mucoperiosteal flap is to be raised is selected.
- *Local anesthesia*—anesthesia to the area is accomplished by block injection to the area and with local infiltration

with a 2% solution of local anesthetic along with vasoconstrictor.

- *Incision lines*—with a scalpel, press firmly against the cortical bone, outline the flap using the periosteal elevator and strip the tissue from the bone.
- *Drilling by bur*—using a small bone bur in low speed drill, make a small square window through cortical plate; using a water spray as a coolant.
- *Removal of cortical plate*—remove the cortical plate of bone and with curette, obtain deep sample of the underlying lesion and place the sample in 10% formalin.
- *Closing of flap*—the surgical site is closed by replacing the flap.

Punch Biopsy

It is rarely necessary in the oral cavity as most of the oral lesions are easily accessible. With this technique the surgical defect that is produced is small and does not require suturing.

In this technique, a sharpened hollow tube (Fig. 8-5); several millimeters in diameter is rotated until underlying bone or muscle is reached. The tissue is then removed in the same manner as in incisional or excisional biopsy.



Fig. 8-5: Different types of punch of different diameter are used in punch biopsy.

Frozen Section Biopsy

It is performed in order to get an immediate histological report of a lesion. It is done to determine whether a lesion is malignant or not. It is also used to evaluate the margins of an excised cancer, to ascertain that the entire lesion is removed at the time of surgery.

The tissue is obtained from lesion and it is kept in deep freeze and then frozen tissue is sectioned and stained to get a prompt diagnosis. In this type of biopsy, the slides cannot be preserved for future reference.

Exfoliative Cytology

In this, the surface of the lesion is either wiped with some sponge material or scraped to make a smear. It is the

microscopic examination of the cells shed from an epithelium. The appreciation of the fact that some cancer cells are so typical that they can be recognized individually has allowed the development of this diagnostic technique, which is developed by Dr. George Papanicolaou Who is also known as the 'father of cytology' and the technique is called as *Pap smear* (Fig. 8-6).

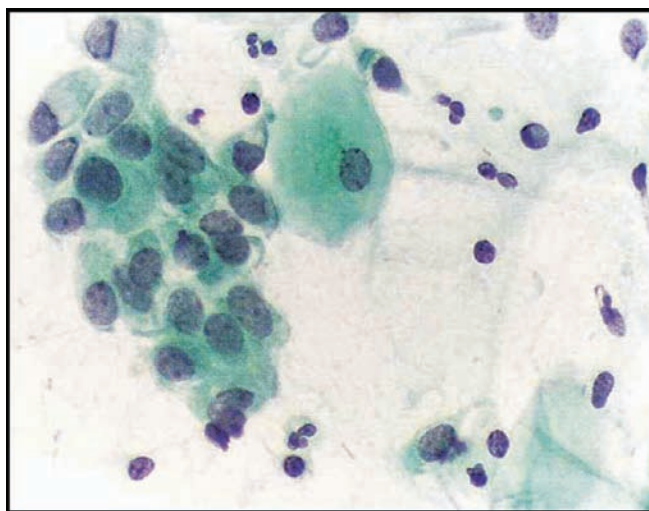


Fig. 8-6: Pap smear (Courtesy Dr Akshay Dhoble).

Principle of Pap Smear

- *Normal cells*—individual cells can often be diagnosed as such microscopically by their large size, their pleomorphism, increased nucleocytoplasmic ratio, hyperchromatism and prominence of nuclei and their abnormal mitosis.
- *Dysplastic cells*—cancer cells exfoliate more easily than normal cells most likely due to their lowered cohesiveness as a result of either decrease in number of tight junctions or lower calcium content.

Advantages

- *Time saving*—cytology is quick procedure so it saves lot of time. It is simple procedure.
- *Painless*—as it is painless, it causes minimum discomfort to the patient.
- *Low cost*—cost of performing cytology is less as compared to cost of biopsy.
- *No anesthesia*—it does not require anesthesia.
- *False negative biopsy*—it helps to check against false negative biopsy.
- *Follow up*—it is especially helpful in a follow up detection of recurrent carcinoma.
- *Screening test*—it is valuable for screening lesions whose gross appearance is such that biopsy is not warranted.
- *Safety*—it is a safe procedure as complications are rare.
- *Rapid diagnosis*—it enables a rapid diagnosis.

- *Bloodless procedure*—as it is bloodless procedure there is less risk of delayed wound healing and infection.
- *Accuracy*—there are report of 100% accuracy in lymph node aspiration from metastatic carcinoma, melanoma, Hodgkin's and Non-Hodgkin's lymphoma.

Disadvantages

- *Firm tumors*—firm tumors may prevent a proper cytodiagnosis due to paucity of cells in the aspirate.
- *False negative results*—oral cytology can give false –ve findings due to inadequate sampling. Another reason for this is many lesion have a thick keratinized surface layer and contain subtle change of dysplasia.
- *Non-assessment*—some specimens cannot be assessed due to poor cellularity.

Indications

- *Patient preference*—as a compromise, when the patient refuses for biopsy.
- *Follow-up*—as a means of follow-up for recurrence in patients who had radiation therapy for the lesion that was superficial or adjacent to bone, periodic recall of high risk patient.
- *Debilitated patients*—in place of biopsy, when dealing with extremely debilitated patients possessing problems to determine a suitable biopsy site.
- *Adjunct test*—as an aid to the diagnosis of some dermatological diseases such as Pemphigus, White sponge nevus, oral malignant and pre-malignant lesions.
- *Periodic review*—periodic review of oral lesion like leukoplakia, lichen planus should be carried out by this method.
- *Rapid evaluation*—for rapid evaluation of an oral lesion that on clinical grounds, is thought to be malignant or pre-malignant and for which, the dentist is unable to obtain permission for a biopsy.
- *Sequential laboratory evaluation*—for sequential laboratory evaluation of an area of the mucosa that has previously been treated by radiation or by excisional biopsies to remove malignancy.
- *Vesicular lesion*—for evaluation of vesicular lesions where facilities for rapid evaluation of Tzanck smears are not available.
- *Population screening*—when population screening is done for the detection of oral cancer exfoliative cytology is the recommended method.

Instruments Used

Instrument which are used in exfoliative cytology are glass microscopic slide, lead pencil, cement spatula or wax carver, wooden tongue depressor, toothpick, canister of cytospray and 95% isopropyl alcohol or ethyl alcohol.

Procedure

- *Information to written on slide*—use of two slides for each site to be sampled. With lead pencil print the patient's name, date when the slide is prepared and the site of the lesion on frosted end of glass microscopic slide.
- *Instrument*—the instrument selected to remove the superficial cell must have a square edge with a contour sufficient to scrape off the superficial layer of cells. When the lesion is very small, the edge of toothpick is effective.
- *Clearing of surface*—clear the surface of oral lesions with debris and mucus.
- *Scraping the tissue*—while the tissue is stretched, the squared edge of the collection instrument is positioned at the back of the lesion and is firmly held and brought forward and pressure applied until visible material is collected. Vigorous scraping of the entire surface of the lesion several times is done with a metal cement spatula or a moistened tongue blade.
- *Spreading of material on microscopic slide*—collected material is then quickly spread evenly over the microscopic slide.
- *Fixing of tissue*—fix it in commercial preparation such as spraycyte, 95% alcohol or equal part of alcohol and ether, immediately before it dries. Then allow it to stand for thirty minutes so that it air dried.
- *Repeat*—repeat the procedure and prepare a second smear.

Interpretation

It is reported by a cytologist as follows into one of the following five classes:

- *Class I (normal)*—it indicates that only normal cells are observed.
- *Class II (atypical)*—presence of minor atypia but no evidence of malignant changes.
- *Class III (indeterminate)*—this is a stage in between that of class II and IV and separates non-cancer cells from cancer cells displaying wider atypia that may be suggestive of cancer but they are not clear-cut and may represent pre-cancerous lesion or carcinoma *in situ* and a biopsy is recommended in such cases.
- *Class IV (suggestive of cancer)*—few cells with malignant characteristic or many cells with borderline features. Biopsy is mandatory in such cases.
- *Class V (positive of cancer)*—cells that are obviously malignant. Biopsy is mandatory in such cases.

Fine-Needle Aspiration Cytology

It is the microscopic examination of an aspirate obtained by inserting a fine needle into the lesion. It is a painless

and a safe procedure for rapid diagnosis. First discovered by Kun in 1847 and reintroduced in 1930 by Martin and Ellis.

Indications

- *Salivary gland pathology*—FNAC of salivary glands is a useful procedure for evaluation of salivary gland tumors.
- *As replacement for extensive biopsy*—it is indicated in lesions in which open biopsy require extensive procedures.
- *Suspicious lymph nodes*—for examination of enlarged clinically suspicious lymph nodes.
- *Recurrence*—to check against recurrence or local extension.
- *Metastatic lesion*—detection of metastatic sq. cell carcinoma within cervical nodes.

Procedure

- *Needle positioning*—first you have to position the needle within the target tissue.
- *Application of negative pressure*—plunger is pulled to apply negative pressure. Needle is moved back and forth within the target tissue to obtain a greater field.
- *Releasing of negative pressure*—negative pressure is then released while the needle remains within the target tissue.
- *Withdrawing the needle*—needle is withdrawn and then the defumed air drawn in the syringe and the aspirate is blown onto the slide.
- *Fixing*—fixing is done in 95% alcohol for 1 hour for Pap stain and a little prolonged for HE stain.

ORALCDX Test

It is highly specialized computer assists analysis of an oral brush biopsy performed on oral tissue. It is the most recent development in oral biopsy technique. This technique is ideal for determining the need for scalpel biopsy in benign-appearing oral mucosal leukoplakia.

Procedure

- *Collection of sample cells*—this technique utilizes a disposable brush to collect a transepithelial sampling of cells (**Fig. 8-7**).
- *Computer screening*—the sample is screened by computer which is programmed to detect cytologic changes associated with premalignancy and squamous cell carcinoma. The computer consists of neural network based image processing system specially designed to detect oral epithelial precancerous and cancerous cells.

- *Image processor*—this is specially designed to detect as few as two abnormal epithelial cells scattered among more than thousand cells on each biopsy specimen.

Interpretation

The specimen is reviewed by a pathologist for final diagnosis.

- *Negative results*—it indicated no epithelial abnormality.
- *Positive results*—it indicated definitive cellular evidence of epithelial dysplasia. Patient should be referred to scalpel biopsy.
- *Atypical results*—it indicates abnormal epithelial changes. This should be referred to scalpel biopsy.



Fig. 8-7: Use of disposable brush to collect sampling of cells in oral brush biopsy.

Molecular Methods

Quantification of Nuclear DNA Content

Principle

The DNA content of nucleus is dependent upon the number of chromosomes. In case of epithelial dysplasia and malignancy there can be polyploidy (more number of chromosomes) or aneuploidy (abnormal number of chromosome). So quantitative analysis of DNA content reflects the total chromosomal content.

Procedure

It is done by flow cytometer analysis. Flow cytometer is automated, precise, reproducible, reliable and objective measuring device of cellular DNA content. Limitation of flow cytometer analysis is that it scans only severe dysplasia of detectable oral premalignancy.

Tumors Markers

Tumor markers are defined as biochemical substances synthesized and released by cancer cells. Tumor markers may be produced by host in response to cancerous substances. Tumors markers can be seen in blood circulation, body cavity fluids, cell membrane and cell cytoplasm.

In oral cavity no markers are found there in cases of oral cancer. In some cases of leukoplakia tumor marker like PCNA, Ki-67 are found which can be value predicting lesion behavior.

Microsatellite Markers

It is one of sensitive method for studying clonal changes in tumors and premalignant lesion. This requires small quantities of DNA. It is done by polymerase chain based Microsatellite analysis.

In oral premalignant lesion loss of heterozygosity is seen.

Immunofluorescence Procedure

Fluorescent dyes such as fluorescein isothiocyanate (FITC) and rhodamine can be chemically linked (conjugated) with antibody globulin without destroying the specificity of the antibody. Such fluorescent labeled antibodies used to detect specific antigen-antibody reaction can be used to locate either antigens or antibodies of known specificity in tissue sections. When tissue sections labeled in this fashion are illuminated with ultraviolet light in an ultraviolet microscope, specific labeled tissue component can be identified by their bright apple green fluorescence against a dark or counter stained background. The technique is used to identify a number of tissue structures and abnormal deposits of antibody globulin and other macromolecules as well as bacteria and viruses in infected tissues and smears.

It is carried out in the following three ways.

Direct Immunofluorescence

- *Application of fluorescent labeled antiserum*—fluorescent labeled antiserum directed against a particular tissue component is applied directly to a thin, unfixed smear or tissue section mounted on a slide.
- *Incubation of slide*—the slide is incubated at 37°C to allow the antigen and the labeled antibody to react.
- *Washing of slide in buffered normal saline*—following incubation, the slide is washed in buffered normal saline to remove un-reacted labeled antibody.
- *Examination under ultraviolet microscope*—the slide is examined in ultraviolet microscope.

Indirect Immunofluorescence

- *Application of unlabeled specific antiserum and FITC conjugated anti-globulin antiserum*—it is directed against a particular tissue component. Applied directly to the smear or tissue section, allowed to react and followed by a FITC conjugated anti-globulin antiserum.
- *Examination under ultraviolet microscope*—following incubation and washing to remove unreacted reagent, the slide is examined in the ultraviolet microscope.
- *Advantages*
 - *Brighter fluorescence*—the fluorescence is brighter because several fluorescent anti-globulin molecules bind onto each of the antibody molecules in the specific antiserum (Fig. 8-8).
 - *Cost effective*—because the process of conjugation is lengthy, there is considerable cost saving and versatility to the indirect technique, which requires only one labelled antiserum.
 - *More staining*—staining of more than one tissue component per slide can also be accomplished.

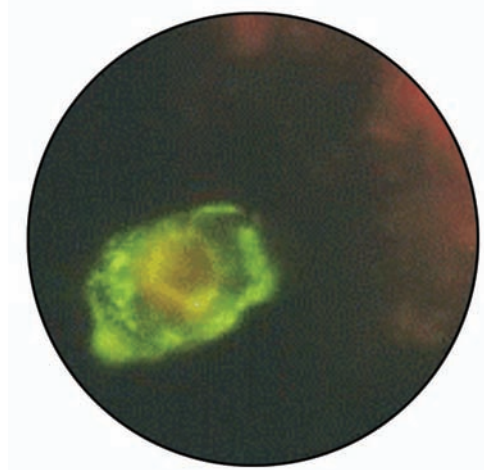


Fig. 8-8: Positive immunofluorescence in case of HSV infection.

Sandwich Technique of Immunofluorescence

- *Why called sandwich*—the name sandwich technique refers to the fact that antigen is sandwiched between two layers of the same specific antibody, one labeled and one not.
- *Reaction of solution of antigen with antibody*—appropriately fixed tissue sections are reacted with a solution of the antigen for which specific antibody is to be identified in the section.
- *Application of FITC labeled antiserum*—following incubation and washing, FITC labeled antiserum with the same specificity as that to be identified in the section, is applied to the section. The labeled antiserum again identifies the location of the tissue component.

Caries Activity Tests (Fig. 8-9)

Caries activity can be defined as the occurrence and rate at which teeth are destroyed by the acid produced by plaque bacteria or it can also be defined as the sum total of new carious lesions and the enlargement of existing carious cavities during the given time.

Uses

- *Preventive measures*—to determine the need and extent of preventive measures.
- *Success of therapeutic measure*—to determine the success of therapeutic measures.
- *Diet counseling and oral hygiene procedure*—to motivate and monitor the effect of education programs related to diet counseling and oral hygiene procedures.
- *Identification of high risk group*—to identify high risk groups and individuals.

Lactobacillus Count Test

It was introduced by Hadley in 1933. It estimates the number of bacteria in the patient's saliva by counting the number

of colonies appearing on tomato Peptone Agar or L.B.B. Agar.

Although it is quick and easy, the results are not available for several days and counting colonies is a tedious and complex process.

Technique

- *Collection of saliva*—stimulated saliva is collected before breakfast by chewing paraffin.
- *Spreading on agar plate*—this is shaken and 1:10 and 1:100 dilutions are spread on the surface of agar plate.
- *Incubation*—these are incubated at 37°C for a period of 3-4 days. The number of colonies is then counted in a Quebec counter.
- *Results*—the count expressed as the average number of colonies per milliliter of the original saliva sample.

Interpretation

- *Immune*—if the count is less than 10^3 then the patient is immune.
- *Slight*—if the count is between 10^3 —5000, caries activity is slight.
- *Medium*—if the count is between 5000— 10^4 , caries activity is medium.
- *High*—if it is more than 10^4 then caries activity is high.

Snyder Test

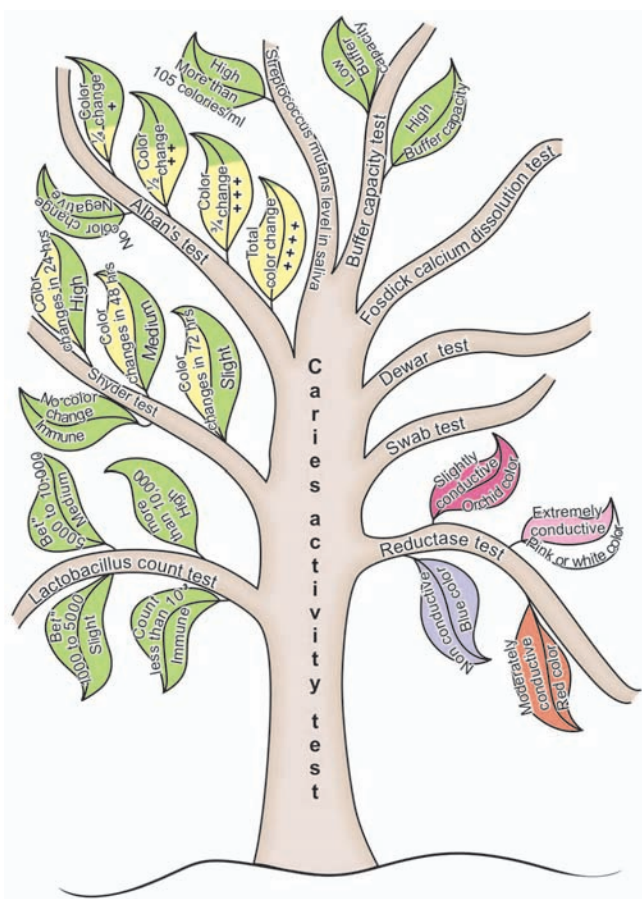
This test measures the ability of microorganisms in saliva responsible for formation of acids from carbohydrate media.

Technique

- *Collection of saliva*—the saliva sample is collected in a manner similar to the Lactobacillus colony count test.
- *Snyder media*—the media contains bactopectone (20 gm), dextrose (20 gm), sodium chloride (5 gm), agar (16 gm) and bromocresol green (0.02 gm).
- *Mixing of media with saliva*—then 0.2 cc of saliva is pipette into the media which is incubated at 37°C for a period of 72 hours.
- *Results*—bromocresol green, being an indicator, changes the color from blue green to yellow in the range of pH of 5.4-3.8. The color change is then correlated with the caries activity.

Interpretation

- *High*—if color changes in 24 hours, caries activity are high.
- *Medium*—if the color changes in 48 hours, it is medium.
- *Slight*—if color changes in 72 hours, it is slight.
- *Immune*—if there are no color change patient is immune to caries.



Alban's Test

It is a modification of Snyder's test. It uses less quantity of agar i.e. 5 ml per tube. Because of its simplicity and its low cost it is recommended for all patients prone to caries.

The main feature of Alban's test is the use of a softer medium that permits the diffusion of saliva and acids without the necessity of melting the medium and use of a simpler sampling procedure in which the patient expectorates directly into the tubes that contain the medium.

Technique

- *Preparation of Alban medium*—60 gm of Snyder test agar is placed in 1 liter of water and the suspension is brought to boil over a low flame or a hot plate at medium heat (excessive heating should be avoided to prevent scorching of medium). When thoroughly melted, agar is distributed, using about 5 ml per tube. The tube should be autoclaved for 15 minutes, allowed to cool and stored in a refrigerator.
- *Mixing and incubation of saliva with Alban medium*—two tubes of Alban medium are taken from the refrigerator and saliva is drooled directly into the tubes and tubes are incubated for 4 days at 37° Celsius.
- *Results*—the tubes are observed daily for change in color. The color change is noted from bluish green to yellow and the depth to which change has occurred is noted.

Interpretation

- *Negative*—no color changes.
- + beginning of color change (from top of the medium towards bottom).
- ++ one-half color changes.
- +++ three-fourth color changes.
- ++++ total color change.

Streptococcus Mutans Level in Saliva

Saliva samples are obtained by using tongue blades (after air drying the tooth for plaque samples). These are then incubated on M.S.B. agar (Mitis Salivarius Bacitracin Agar). The number of colonies is then used to estimate the caries activity and more than 10^5 colonies per ml of saliva is indicative of high caries activity.

Detection of mutans streptococci in dental plaque and stimulated saliva



Attach the strips to the cap and recap the vial

Incubation (48 hours)

Interpretation (screening strips)

Interpretation (site strips)

Buffer Capacity Test

Technique

- *Collection of saliva*—10 ml of stimulated saliva is collected under oil at least one hour after eating; 4 ml of this is measured into a beaker.
- *Adjusting pH of saliva to 7*—after collecting the pH meter, the pH of saliva is adjusted to 7.0 by addition of lactic acid or base at room temperature.
- *Results*—the level of lactic acid in the graduated cylinder is re-recorded. Lactic acid is then added to the sample until a pH of 6.0 is reached. The number of millimeter of lactic acid needed to reduce pH from 7.0 to 6.0 is a measure of buffer capacity of saliva.

Interpretation

- *Low buffer capacity*—saliva sample requiring less than 0.45 ml of standard hydrochloride acid to reduce the pH to 5 has low buffer capacity.
- *High buffering capacity*—saliva sample requiring 0.45 ml or more has high buffering capacity.

Fosdick Calcium Dissolution Test

Technique

- *Collection of saliva*—twenty five milliliters of gum stimulated saliva is collected.
- *Keeping powdered enamel with saliva*—part of this is analyzed for calcium content and the rest is placed in an eight inch sterile test tube with about 0.1 gm of powdered human enamel.
- *Shaking of tube*—the tube is sealed and shaken for four hours at body temperature after which, it is again analyzed for calcium content. If paraffin is used, a concentration of about 5% glucose is added.

Interpretation

- The amount of enamel dissolution increases as the caries activity increases which is indicated by an increase in the calcium content of saliva.

Dewar Test

Technique

- *Collection of plaque sample*—plaque samples are collected from the gingival third of buccal tooth surfaces and placed in Ringer's solution.
- *Mitis Salivarius agar plate*—the sample is shaken until homogenized. The plaque suspension is streaked across a Mitis-Salivarius agar plate.
- *Incubation*—after aerobic incubation at 37° C for 72 hours, the culture is examined under a low power microscope and the total colonies in 10 fields are recorded.

Interpretation

- This test is an attempt to semi-quantitatively screens the dental plaque for a specific group of caries causative organisms including streptococci.

Swab Test

The swab test involves sampling of the oral flora by swabbing the buccal surface of teeth and placing it in Snyder media. This is incubated for 48 hours and the pH changes are read and correlated with caries activity.

Reductase Test

This test measures the activity of salivary enzyme reductase.

Technique

- *Collection of saliva and mixing with diazoresorcinol*—saliva is collected and the sample is mixed with a diazo-resorcinol, which colors the saliva blue.
- *Results*—the change in color from blue to red is measured after 30 seconds and 15 minutes and this is taken as measure of caries activity.

Interpretation

- *Non-conductive*—if it remains blue after 15 minutes it is non-conductive.
- *Slightly conductive*—if it changes to orchid after 15 minutes, it is slightly conductive.
- *Moderately conductive*—if it changes to red after 15 minutes, moderately conductive.
- *Highly conductive*—if it changes immediately to red, it is highly conductive.
- *Extremely conductive*—if it changes to pink or white immediately, it is extremely conductive.

Hematological Investigation

Normal values are given in Table 8.1.

Collection of Blood Specimen

- *Capillary blood specimens*—these are convenient for office and chair side procedures. The specimen is obtained by pricking the patient's finger (an earlobe, ankle pad can also be used). Procedure is as follows
 - *Equipment*—jar of sponge soaked in 70% alcohol.
 - *Application of alcohol to patient finger*—the palmar surface of the patient's index finger or second finger is gently rubbed with sponge soaked in 70% alcohol. Excess alcohol is wiped off and the alcohol is allowed to evaporate.

- *Puncturing the finger with blood lancet*—the prepared area is rapidly punctured with a blood lancet taking care not to squeeze the finger (**Fig. 8-10**).
- *Collection of blood sample*—the patient's hand is at about the waist level and the first drop of the blood that wells up is wiped out with dry sponge. The next drop that appears allows to flow into a capillary tube or is collected on a glass slide.



Fig. 8-10: Collection of capillary blood specimen is done by finger pricking.

- *Venous blood specimen*—venous blood specimens are used for majority of tests that are to be performed in clinical diagnostic laboratory. Venipuncture is usually performed in antecubital vein (**Fig. 8-11**). Procedure is as follows:
 - *Equipment*—a 10 to 20 ml sterile disposable syringe with a 20-gauge needle or Vacutainer. Needle, adaptor and evacuated collection tubes, tourniquet or approximately 20 inch of soft tissue rubber tubing,



Fig. 8-11: Venous blood is taken by venipuncture method.

less than ½ inch in diameter. Jar of sponges soaked in 70% alcohol and jar of dry sponge.

- *Preparation of patient*—prepare the patient and equipment. Apply the tourniquet and select the vein to be punctured.
- *Preparation of puncture site*—prepare the puncture site with 70% alcohol and air dry.
- *Collection of blood*—insert the needle through the skin and into the vein. Aspirate blood into a syringe or Vacutainer tube and remove the tourniquet.
- *Maintenance of puncture site*—place dry cotton swab over the puncture site and withdraw the needle. Bring the patient's forearm back against his upper arm while he maintains finger pressure on the cotton at the puncture site with the other hand.
- *Delivery to laboratory*—specimen should be delivered to the laboratory within 1-2 hours in a tube which should be well sealed and protected from breaking.
- *Arterial blood specimen*—arterial blood specimen is obtained by puncturing the radial artery.

Total Erythrocyte Count

It gives the number of erythrocytes per cubic millimeter in circulating blood and also gives an indirect estimate of hemoglobin in blood. It is determined by hemocytometer.

Procedure

- *Office and chair side*—the test is performed manually under a microscope by direct counting of the number of cells in diluted sample of blood confined in a calibrated chamber of special glass microscope slide (hemocytometer technique).
- *Automated procedure*—the electronic counting of erythrocyte is usually carried out with equipment such as *coulter counter model* (Fig. 8-12). The sample of whole



Fig. 8-12: Hemocytometer is used to calculate RBC count.

blood is aspirated into the machine and automatically diluted with modified Eagle's solution before being drawn through three narrow chambers equipped with electrodes. As the cells pass through an aperture, their presence and dimensions are accurately recorded by a drop in voltage across the electrodes. The reading for three aperture tubes is obtained as average and an average RBC count is displayed and printed out by the machine.

Interpretation

- Earliest signs and symptoms of hematological disease affecting erythrocytes are anemia and polycythemia. It is reduced in some anemia and increased in polycythemia and dehydration.

Erythrocytes Indices

In evaluation of nature of anemia, assistance is obtained by calculating standard indices relating to the size of RBCs. By measuring these indices we can classify anemia as microcytic, macrocytic and normocytic and hypochromic and normochromic (Tables 8.1 and 8.2).

Types

- *MCH*—the hemoglobin content of erythrocyte is referred to as the mean corpuscular hemoglobin (MCH) expressed in picograms of hemoglobin per cell.
- *MCHC*—the concentration of hemoglobin in the erythrocyte is referred to as the mean corpuscular hemoglobin concentration (MCHC)
- *MCV*- average red cell volume referred to as the mean corpuscular volume (MCV) is expressed in cubic microns per cell.

Table 8-1: Different erythrocytes indices

Indices	Calculation
MCH	$\frac{\text{Hemoglobin concentration (g/dl)}}{\text{RBC in million/mm}^3} \times 10$
MCHC	$\frac{\text{Hemoglobin concentration (g/dl)}}{\text{Hematocrit}} \times 100$
MCV	$\frac{\text{Hematocrit}}{\text{RBC in million/mm}^3} \times 10$

Table 8.2: Different types of anemia and erythrocytes indices

Types of anemia	MCV	MCH	MCHC
Microcytic hypochromic	Decreased	Decreased	Decreased
Macrocytic normochromic	Increased	Increased	Normal
Normocytic normochromic	Normal	Normal	Normal

Hematocrit

It is a measure of volume percent of packed red blood cells in whole blood.

Indications

- *Anemia*—the classic signs and symptoms of anemia indicate for the measurement of the hematocrit value.
- *History of blood loss*—history of excessive blood loss, pallor of oral mucous membrane, palate, conjunctiva, nailbeds.
- *Evaluation of patient for general anesthesia*—To evaluate patient's ability to tolerate general anesthesia or oral surgical procedures.
- *Others*—unexplained syncope, excessive fatigue, glossitis and atrophy of lingual papillae, recurrent and persistent gingivitis and stomatitis.

Technique

- *Equipment*—2 heparinized microhematocrit capillary tubes. A box of matches and microhematocrit centrifuge equipped with reader and magnifier.
- *Collection of blood*—blood is obtained by a finger prick or venipuncture and is allowed to flow by capillary action into two microhematocrit tubes, until each is about $\frac{3}{4}$ th filled. The volume of blood should be free from air bubbles.
- *Sealing of tube*—the end of each tube away from the column of blood is sealed by melting the glass in the flame of a match or small gas jet without heating the blood.
- *Keeping tube in centrifuge*—the tubes are cooled and placed opposite to each other in the slots of the head of the centrifuge, with sealed ends pointing outward. The centrifuge head covers are closed and the centrifuge timer is set for 5 minutes.
- *Centrifugation*—the centrifuge is started and it accelerates to 12,500 rpm and stop automatically at the end of required time.
- *Results*—the centrifuge head covers are removed and with the aid of magnifying lens for greater accuracy, the hematocrit is read by means of scale incorporated in the head of the instrument.

Interpretation

- *Increase*—increase in the value indicates primary and secondary polycythemia and dehydration.
- *Decrease*—value is decreased in anemia.

Measurement of Hemoglobin Concentration

It is expressed in gm/dl. It is require to obtain information about circulating RBC's and the oxygen carrying capacity. It is also used for calculation of MCHC and MCH.

Technique

- *Photoelectric calorimeter*—the hemoglobin concentration is usually measured on anti-coagulated venous blood by reacting the sample with a reagent that converts the hemoglobin to stable colored product. The concentration of this colored compound is measured in photoelectric colorimeter in comparison with the standard.
- *Drabkin's method*—different methods used are Drabkin's technique in which hemoglobin is converted into the stable pigment cyanmethemoglobin. Drabkin's solution contains potassium cyanide and ferricyanide and sodium bicarbonate.
- *Other method*—other techniques used are Sahli's method (0.1 N hydrogen chloride), oxyhemoglobin method (0.1 sodium carbonate).

Interpretation

- *Increased*—it is increase in primary and secondary dehydration.
- *Decrease*—hemoglobin concentration decreased in anemia.

WBC Count

In the management of dental patients total WBC count is used as one of the index of presence of systemic infection and to rule out possibility of leukemia and malignant neutropenia (Table 8-3).

The total WBC count can be calculated manually with a hemocytometer or with an automated cell counter. To count WBCs in the presence of RBCs, RBCs are lysed by diluting the blood sample with dilute acetic acid or equivalent reagent supplied by the manufacture of the automated equipment leaving the WBCs intact.

Table 8-3: Interpretation of WBC count

Increased in (leukocytosis)	Decreased in (leukopenia)
• Infection –acute and chronic • Leukemia • Polycythemia • Trauma • Exercise, fear and pain • After general anesthesia	• Aplastic anemia • Influenza, measles and respiratory tract infection • Catarrhal jaundice • Early leukemia • Depression of bone marrow due to certain drugs • Drug and chemical toxicity, shock

Differential Leukocyte Count

As the WBCs in circulating blood are of several types and of varied origin, the total leukocyte count is of limited use without a differential count of the various types of cells present. Five types of WBCs are commonly present in circulating blood. They are neutrophils, lymphocytes, monocytes, basophils and eosinophils.

Technique

- **Staining**—it is performed on air dried smear of blood stained with Wright's or other Romanovsky-type stain to differentiate types of WBCs from each other. Nowadays various instruments are used to calculate DLC count (**Fig. 8-13**)
- **Relative percentage**—the relative percentage of each type of WBC is obtained by counting a minimum of 100 to 200 cells per slide and the fields counted are selected in such a way that the effect of uneven distribution of WBCs in the smear is overcome.



Fig. 8-13: DLC count is calculated by instrument.

Interpretation

It is discussed in Table 8-4.

Platelets

Platelets are small $2\ \mu$ to $5\ \mu$ particles released from the cytoplasm of large multinucleated cells (megakaryocytes) located in the red bone marrow.

Because of the platelet's small size and tendency to aggregate, (an important phenomenon related to their function in the hemostatic mechanism) platelet counts on peripheral blood smear have a large technical error.

Counts are performed manually using a special hemocytometer counting chamber and a phase contrast microscope or by means of an automated cell counter such as the Technicon Autoanalyzer. Interpretation of platelet is discussed in Table 8.5.

Bleeding Time

It measures the time required for hemostatic plug to form. Lack of any clotting factor and platelet abnormalities may increase the bleeding time. It is a useful screening test in patients with a history of prolonged bleeding following previous surgery. It should be carried out in patients in whom certain symptoms are with strongly suggestive of bleeding disorders.

Table 8-4: Interpretation of different types of WBCs in disease

Increased in	Decreased in
Neutrophils	
• Pregnancy, exercise • Infection, inflammation • Neoplasm, severe hemorrhage • Hemolysis and erythroblastosis fetalis • Intoxication by drugs and poisons like corticosteroids	• Aplastic anemia • Cyclic neutropenia • Malignant neutropenia • Drug induced myelosuppression • Early leukemia
Lymphocytes	
• Lymphocytic leukemia • Mumps—German measles • Whooping cough • Chronic infection • Convalescence from acute infection	• Aplastic anemia • Myelogenous anemia
Monocytes	
• Hodgkin disease • Monocytic anemia • Malaria—kala-azar • Sub-acute bacterial endocarditis • Tuberculosis	• Aplastic anemia • Acute leukemia
Eosinophils	
• Parasitic infection • Allergic disease • Drug reaction dermatoses like pemphigus, pemphigoid, atrophic dermatitis • Scarlet fever	• Immune defect • Acute stress • Injection of adreno-corticotrophic hormone • Typhoid fever • Aplastic anemia
Basophils	
• Chronic myelogenous leukemia • Polycythemia • Myelofibrosis	

Table 8.5: Interpretation of platelets

Increased in (Thrombocytosis)	Decreased in (Thrombocytopenia)
• Chronic myelocytic leukemia • Polycythemia vera • Hemolytic anemia • Trauma	• Thrombocytopenic purpura • Immune reaction • Transfusion • Aplastic anemia

Technique Equipment

- **Equipment**—sphygmomanometer, cuff, jar of sponges soaked in 70% alcohol and jar of dry sponges, blood lancet, 3 to 4 pieces of filter paper about 1 inch square, stop watch (**Fig. 8-14**).
- **Patient position**—the patient is seated comfortably with his arm supported on the chair arm or his own thigh.
- **Application of cuff to patient**—the sphygmomanometer cuff is applied to the patient's upper arm and inflated to 40 mm pressure.

- *Scrubbing area with alcohol swab*—an area on the palmar (inner) surface of the forearm about halfway between elbow and wrist that is free of superficial veins, is located and prepared by scrubbing with an alcohol soaked swab.
- *Punctured with blood lancet*—after the area is air dried, the skin of the prepared area is tensed and punctured with a blood lancet cutting deep enough such that the hit is firmly pressed against the skin of the arm.
- *Results*—the lancet is immediately removed and the time noted on a watch equipped with a seconds hand. Every 30 seconds by the watch, an edge of filter paper is touched against the drop of blood that wells up. The length of time until the bleeding ceases is the bleeding time in minutes.
- *Cleaning the area*—the area is finally cleaned with a swab slightly moistened in 70% alcohol.

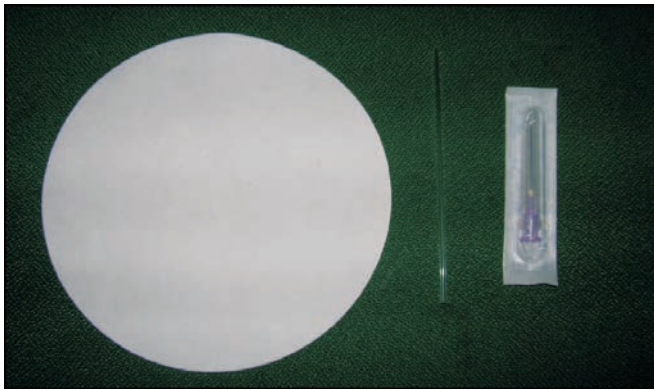


Fig. 8-14: Filter paper, blood lancet are used to calculate bleeding time.

Interpretation

- *An abnormal bleeding time*—it is usually the result of abnormalities in the structure or ability of the capillary blood vessels to contract or abnormalities in the number or functional integrity of the platelets.
- *Contributing factors*—a bleeding time of 5 minutes does not mean that a patient will stop bleeding from any type of wound in 5 minutes. The time for bleeding to stop is related to the way in which the wound is produced, the caliber of the vessels involved in the hemorrhage, the amount to tissue damage adjacent to the wound and systemic factors such as blood pressure and individual response to the type of anesthesia.

Capillary Fragility Test (Tourniquet Test and Rumpel Leede Test)

It is the test of the ability of superficial capillaries of the skin of the forearm and hand to withstand an increased intraluminal pressure and a certain degree of hypoxia. It is

done by occluding veins of the upper arm with a blood pressure cuff for five minutes.

Indications

- *Bleeding abnormalities*—it is used to screen for bleeding abnormalities especially where there is suspicion of platelet disorders.
- *Petechiae in oral cavity*—the discovery of petechiae in the oral cavity or on the skin is the most common indication for the capillary fragility test; especially where the petechiae are confined to the oral cavity and could also conceivably have resulted from local trauma or denture irritation.
- *Scurvy*—in dental practice, it is used as a screening test for scurvy, which is an etiological factor in periodontal disease.

Technique

- *Equipment*—sphygmomanometer and cuff, microscope slide.
- *Patient position*—the patient is seated with one arm supported on the chair arm or thigh.
- *Instruction to patient*—the dentist should explain the patient that his blood pressure will be taken and the cuff will be left on a little longer than the patient may be used to. He should be further explained that the procedure may make the patient's arm a little numb and even painful, but that discomfort will quickly disappear once the cuff is removed.
- *Examination of hand of patient*—during this time, the dentist examines the patient's forearms and hands for any petechiae and records their location with an ink spot. The blood in a petechial hemorrhage lies extravascularly and should not disappear upon pressure with the surface of a glass microscope slide.
- *Application of sphygmomanometer*—the sphygmomanometer is applied customarily and the patient's systolic and diastolic arterial blood pressure is recorded.
- *Inflation of cuff*—the pressure in the cuff is reduced to zero and the cuff is reinflated to a point halfway between systolic and diastolic pressure. This pressure is maintained for 5 minutes during which the forearm and hands are examined for development of new petechiae. The patient should not be moved vigorously while the cuff is in place because movement will increase anaerobic muscular glycolysis, lactate accumulation and thus result in pain.
- *Results*—after 5 minutes, the cuff is removed and the patient is allowed to exercise his arm to restore the circulation in the arm. The blood vessels of normal individuals will withstand these conditions and

petechial hemorrhages will not appear on the forearm and hand from rupture of superficial capillaries.

- **Positive test**—if any unequivocal petechiae develop distal to the cuff (toward the hand) after cuff has been applied, the test is recorded as positive (**Fig. 8-15**).
- **Negative test**—if only one or two petechiae are seen or if those seen are not convincing in their appearance, a negative test is recorded; retesting is done if required by investigating dentist.



Fig. 8-15: Appearance of petechiae on hand after tourniquet test.

Clotting Time

Procedure

- **Feeling of blood in dry tubes**—one millimeter of venous blood is placed in each of the four dry tubes of standard size, maintained in a water bath.
- **Tilting of tube**—the first tube is tilted at 30 seconds interval, until the blood no longer flow. The next tube it tilted until clotting occurs, after which the third and fourth tubes are similarly treated.
- **Results**—the average time between venipuncture and clotting in the last three tubes is expressed in minutes as the clotting time. The normal range is 10 to 25 minutes.

Interpretation

- **Increases**—it is prolonged in diseases affecting stage II and stage IV of coagulation. It is also increased in cirrhosis, hemophilia A and B, factor XI deficiency, hypofibrinogenemia and heparin and dicumarol anticoagulant therapy.

Erythrocyte Sedimentation Rate (ESR)

It is a measure of the rate at which RBCs sediment in a tube of plasma. The test is helpful in following the progress of some chronic infections (tuberculosis and osteomyelitis)

as well as diseases characterized by altered globulins such as the collagen diseases, nephritis, rheumatic fever and dysproteinemias.

Technique

- **Mechanism**—the rate is accelerated when there are changes in the physicochemical properties of plasma or the red cell surface or when the changes in plasma protein cause the RBCs to aggregate.
- **Westergren method**—in the Westergren method, a graduated sedimentation tube is filled with oxalated blood and placed in an absolutely vertical position (**Fig. 8-16**).
- **Results**—the erythrocyte level is read at 10 minutes intervals and at the end of an hour.

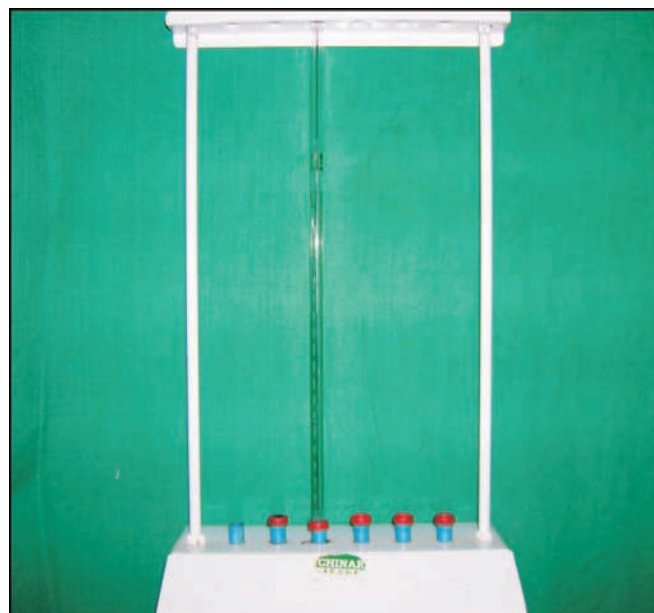


Fig. 8-16: ESR is calculated by Westergren method.

Hematological Investigation not so Frequently Required in Dental Office

- **Partial thromboplastin time**—it is the time, in seconds, that is required for a clot to form in a sample of oxalated plasma, to which a partial thromboplastin reagent and calcium chloride is added.
- **Prothrombin time**—it is the time in seconds that is required for fibrin threads to form in citrated or oxalated plasma, where known amount of tissue thromboplastin and calcium is added. Since last two reagents are present in excess, any delay in clotting in this test is suggestive of an abnormality of the prothrombin complex or a very severe fibrinogen deficiency.
- **Schilling test**—several types of megaloblastic macrocytic anemias of different etiologies are caused by vitamin B₁₂ deficiency. Schilling test is a measurement of patient's

ability to absorb orally administered radioactive vitamin B₁₂ labeled with ⁶⁰Co. Following oral administration of the radioactive vitamin B₁₂, unlabeled vitamin is given intramuscularly, as a flushing dose to induce urinary excretion of the labeled vitamin, which is measured in a 24 hour urine specimen. Patients with pernicious anemia (who are unable to absorb orally administered vitamin B₁₂) excrete less than 5% of the orally administered dose in comparison with excretion of 8 to 25% by normal individuals.

- *Serum iron and total iron binding capacity*—iron deficiency is the most common cause of anemia; however anemia occurs only after depletion of iron stores and iron deficiency may exist for some time before the appearance of anemia. Iron deficiency is usually detected on the basis of the amount of iron bound to transferrin in the plasma (serum iron) and the total amount of iron that can be bound to the plasma transferrin *in vitro* (total iron binding capacity or TIBC).
- *Bone marrow aspiration*—detailed evaluation of patients with anemia, leukemia, multiple myeloma, metastatic malignancy and other space occupying lesions frequently involves study of the immature erythropoietic and leukopoietic cells of the bone marrow. This is accomplished by aspiration of red bone marrow cells through a large bore needle inserted through a trochar into the iliac crest or sternal marrow space. The aspirate is then smeared thinly on a glass slide or coverslip, stained with Wright's stain and examined in the same ways as a smear of capillary or venous blood.
- *Bone marrow biopsy*—bone marrow biopsy is also used where an indication of the spatial relationship and degree of hyperplasia or hypoplasia of the cellular element in the marrow is needed.
- *Platelet survival time*—it is measured in thrombocytopenic purpura and other diseases in which rapid destruction of platelet occurs. A sample of autologous or isologous platelets labeled with radioactive ⁵¹Cr is added to a sample of patients blood and re-injected intravenously. The amount of radioactivity is then measured in blood samples taken at intervals over the next few days, depending on the design of the experiment. The normal platelet survival time (time required for the circulating radioactivity to drop to 10% of its peak value after injection of ⁵¹Cr labeled platelet) is between 8 to 9 days.

Blood Chemistry

Detection of Diabetes Mellitus

Diabetes mellitus is a disease of insidious onset that is not infrequently complicated by serious tissue changes leading

to permanent cardiovascular, renal, cerebral and optic damage.

Importance of Detection of Diabetes Mellitus in Dentistry

- *Periodontal therapy*—the response of diabetic patients to periodontal therapy may be much less satisfactory than non-diabetic, under similar conditions.
- *Healing of oral tissue*—the healing of oral tissues following surgery in diabetics is slower. Complication like tissue necrosis and secondary infections may occur in diabetic patient.
- *Oral disease*—certain oral diseases are predisposed to occur in association with diabetes mellitus (e.g. thrush, denture sore mouth).
- *Oral infection*—the systemic effects of acute localized oral infections may be much greater in a diabetic than in a non-diabetic.

Indications

- *Evaluation of patient suspected of diabetes mellitus*—for evaluation of a patient suspected of having diabetes mellitus. Its presence should be suspected whenever any of the following are found: history of weight loss in the presence of adequate diet, history of excessive thirst and attendant excessive and frequent urination, history of repeated episodes of boils, skin infections and periodontal abscesses and presence of severe periodontitis with excessive bone loss.
- *Screening test*—as a screening test for diabetes mellitus, where the dentist suspects the occurrence of disease in the absence of characteristic signs and symptoms.
- *Family history*—testing of the patient with a family history of diabetes.
- *Known diabetes patient*—as a measure of degree of control of disease in a patient who is known to be a diabetic but who is not under regular medical care and is unwilling to accept referral to a physician for reevaluation of the disease.

Types

- *Urine glucose estimation*—many diabetics excrete glucose in their urine, testing the urine with commercial available reagent strips (Tes-Tape, Clinistix, Chemstrip) is the most convenient screening method of diabetes (Fig. 8-17). But in patient with mild diabetes urine may not exhibit glycosuria. False positive urine sugar tests also occur in individuals with renal glycosuria.
- *Blood glucose estimation*—this is most accurate method for detection of diabetes mellitus. Three schedules for determination of blood glucose concentration are used:

- *Fasting blood glucose*—determination of fasting blood glucose is frequently used as a screening test for hyperglycemia in hospitalized patients. On an outpatient basis, however, maintenance of the fasting state from the evening meal until the next morning is difficult to ensure and may lead to a degree of hypoglycemia that an outpatient cannot tolerate without fainting.
- *Two hours postprandial blood glucose*—determination of the blood glucose two hours after a meal containing 75 gm of carbohydrate (2-hours postprandial blood glucose) is a more sensitive measurement of the hyperglycemia associated with diabetes. Both fasting and 2-hour postprandial blood glucose measurement can be carried out on capillary blood using the Dextrostix, Visidex, or Chemstrip reagent strip. Due to its convenience, simplicity and sensitivity, this test is recommended as the best screening procedure for a dentist to use if he suspects that a patient may be a diabetic and feels that he must explore the problems further by himself prior to medical referral.
- *Oral glucose tolerance test*—glucose tolerance test is one of the procedures used for the definitive diagnosis of diabetes mellitus. It is performed on series of blood samples in a laboratory and is used to confirm the diagnosis of a diabetic patient who does not exhibit constantly elevated fasting blood glucose.
- *Equipment*—blood lancet, jar of sponges soaked in 70% alcohol and jar of dry sponges. Dextrostix, Visidex, or Chemstrip bG reagent strip, wash bottle, urine jar, reagent strip to test for glycosuria.
- *Collection of blood sample*—blood sample is taken with the help blood lancet.
- *Application of reagent strip to blood*—the first drop that wells up is wiped away and the next few drops are freely applied to the entire reagent area on the printed side of blood glucose reagent strip. The blood is allowed to permeate and react with the reagents contained in the strip for exactly 1 minute.
- *Removal of blood from strip*—the blood is then washed from the strip as completely as possible with a sharp stream of water from the wash bottle or faucet.
- *Results*—the test area of the strip is immediately compared with the color chart on the side of the bottle, matching the strip; if the color obtained is intermediate between two color blocks, the result is inappropriate, otherwise, the value for a matching block is read directly.
- *Urine glucose*—the patient is requested to void a sample of urine in the specimen jar provided. This is tested for the presence of glucose by using the appropriate reagent strip.
- *Blood glucose*—the normal range of blood glucose in the fasting state is approximately 70 to 110 mg/dl. The concentration rises to about 160 mg/dl following a meal in normal persons but returns to the fasting level within 2 hours. In diabetics of moderate severity, fasting levels may reach 200 mg/dl, postprandial levels are greater than this and may persist for longer than 2 hours after meals. The diagnosis of diabetes mellitus should be made only after a complete medical history, physical examination and appropriate laboratory tests carried out by a physician. If the 2 hour postprandial blood glucose is above 140mg/dl, the patient is clearly metabolizing glucose in an abnormal fashion.
- *Urine glucose*—when the blood level of glucose exceeds 160 to 180 mg/dl, the normal renal threshold for glucose is exceeded and so glucose will appear in urine which will give a positive test for glucose. The renal threshold for glucose may become elevated as a result of kidney disease and this phenomenon is an important cause of false negative tests for glucosuria in diabetes mellitus and number of other diseases which are associated with hyperglycemia.



Fig. 8-17: Glucometer used for detection of blood glucose.

Technique

Two hour postprandial blood glucose

- *Diet instructions*—patient may take serving of 1 fruit juice plus 1 bowl of cereals with 2 tea spoonful of sugar plus 1 cup of milk plus coffee or tea with sugar plus anything he desires at breakfast at 8 am. If the patient is to be retested in the afternoon, he may eat 1 sandwich plus 1 piece of cake or pie plus coffee or tea with sugar or milk plus anything else he wishes for lunch at 1 pm.

Oral glucose tolerance test

- *Significance*—for many years, glucose tolerance test has been accepted procedure for making a definitive diagnosis of diabetes mellitus and for distinguishing diabetes from other causes of hyperglycemia such as hyperthyroidism. Elevated levels for fasting plasma

glucose, in this context include any concentrations equal to or greater than 140 mg/dl. The OGTT should be administered only to healthy ambulatory patients who are known to be taking no drugs that interfere with the laboratory determination of glucose.

- *Oral glucose challenge*—the oral glucose challenge traditionally used in the glucose tolerance test has varied from 50 to 100 gm, administered either as a glucose solution, and commercially prepared carbohydrate load or a meal equivalent to this glucose dose.
- *Subject preparation*—the test should be performed in morning, after at least 3 days of unrestricted diet (greater than 150 gm of carbohydrate daily) and physical activity. The subject is made to fast for at least 10 hours but not more than 16 hours before the test (water is permitted during this period). The subject should remain seated and should not smoke throughout the 2 hours of test period.
- *Fasting blood sample*—a fasting blood sample should be collected.
- *Sample after oral glucose challenge*—subject has to drink the glucose dose in a concentration; no greater than 25 gm/dl of flavored water, in about 5 minutes. Zero time is the beginning of the drink and blood samples are then collected at 30 minutes interval for 2 hours (a total of five blood samples per test).
- *Storage of sample*—capillary blood samples are preferred. Blood sample should be collected in a tube containing sodium fluoride to inhibit glycolysis and potassium oxalate to prevent clotting. The sample should be centrifuged and separated within 4 hours and plasma freezes unless glucose levels are to be determined immediately.

Blood Chemistry Investigation not so Frequently Carried Out in Dental Office

- *Serum calcium, phosphorus*—it is indicated when you suspect Paget's disease, fibrous dysplasia, primary and secondary hyperparathyroidism, osteoporosis, multiple myeloma, osteogenic sarcoma or metastatic malignancy. It acts as initial screening procedures. The concentration of calcium in serum and body fluids tends to vary inversely with the concentration of inorganic phosphorus (2 mg/dl to 5 mg/dl). The product of serum calcium concentration and serum phosphorus concentration is constant at about 30 to 40 in normal adults, but may be as high as 50 to 60 in growing children. At serum calcium below 7 mg/dl (such as may occur in patients with hypoparathyroidism), signs of tetany (neuromuscular excitability, positive Chvostek's sign) appear.
- *Serum alkaline phosphatase*—alkaline phosphatase occurs in many tissues of the body, but notably in osteoblasts.

Increase in serum concentration of this enzyme is seen primarily as a result of increased osteoblastic activity but also in association with obstructive liver diseases and a variety of miscellaneous conditions such as malignancy or abscess of the liver, amyloid disease, leukemia and sarcoidosis. It should be remembered that increased osteoblastic activity is not restricted to sclerosing (radiopaque) bone lesions but may be quite high in lytic (radiolucent) bone lesions also, as a result of remodeling of the surrounding bone that accompanies a lytic lesion.

- *Serum uric acid*—concentration usually lies in the range of 4 mg/dl to 8.5 mg/dl for males and 2.8 mg/dl to 7.5 mg/dl for females. Uric acid is a metabolic end product of nucleoprotein metabolism derived from the purine molecule. Increase occurs in acute phases of diseases such as leukemia, lymphomas, anemia or lobar pneumonia; in which there is rapid destruction of large number of DNA rich leukocytes. Measurement of uric acid is important in the evaluation of intrinsic disease of temporomandibular joint, particularly when nodules consistent with gouty tophi are noted about the face or ears. In gout, values of 8 mg/dl to 15 mg/dl for serum uric acid are usually observed.
- *Serum albumin and globulin*—the concentrations of serum albumin and globulin, as well as the total serum protein are routinely measured as part of the standard blood chemistry profile.
- *Serum protein electrophoresis*—it is useful screening test in patients with suspected oral lesions of multiple myeloma or systemic lupus erythematosus. It is usually indicated when radiolucent defect detected in the jaw. In serum protein electrophoresis a small volume of serum is subjected to a low voltage electric current causing the proteins of the serum to migrate at different rates. When the serum proteins are distributed across a piece of filter paper or a block of starch or other gelled supporting medium, they may be separated and stained as relatively distinct bands and the concentration of each serum protein component calculated.
- *Serum bilirubin*—jaundice can be recognized by examination of the color of skin, oral mucous membrane and sclera of eyes. It may indicate the presence of hepatitis which can constitute an infectious hazard for a dentist and his patients. Jaundice is usually not evident until total serum bilirubin (derived from red blood cells broken down at the end of their normal 120 days life-span) rises from a normal range of 0.1 mg/dl to 0.8 mg/dl to above 2 mg/dl to 3 mg/dl.
- *Thyroid function test*—it is currently measured either by uptake of a radioisotope by the thyroid gland or by direct measurement of thyroid hormones, triiodothyronine (T_3) and thyroxine (T_4) in serum. T_4 levels in serum are

Table 8-6: Chart of normal values

Test	Normal value
Hemoglobin	Males—14- 17.4 gm/dl Females—12.3 to 15.3 gm/dl
Red blood cell	Males—4.5 –5.9 million/mm ³ Females—4.5 –5.1 million/mm ³
WBC	4000-11000 / mm ³
Platelet count	1,50,000 –4,50,000 /mm ³
Hematocrit	At birth—54 Adult males—42-50% Adult females—36-45%
Red cell indices MCV MCH MCHC	80-96 cu micron 27-31 pg 32-36 gm/dl
ESR	Males (under 50) <15mm/hr Males (over 50) < 20 mm /hr Females (under 50) < 20 mm/hr Females (over 50) < 30 mm / hr
Differential WBC count (range) Neutrophils—50-77 % Eosinophils—0-4 % Basophils—0-2 %	Absolute value 2500-7000/ul 50-500/ul 0-100/ul
Lymphocytes—20-47% Monocytes—0-9 %	1000-4000/ul 200-800/ul
Reticulocyte count	0.5-2.5 %
Bleeding time	1-6 minutes
Clotting time (capillary) (Lee and White)	3-5 minutes 4-10 minutes
Clot retraction	½ of original mass in 1 hr
Prothrombin time	12-15 seconds
Thrombin time	10-12 seconds
Plasma fibrinogen	150-400 mg/dl
PTTK (kaolin activation)	30-35 seconds

Table 8-7: Normal blood chemistry value

Test	Material	Normal value
Amylase	Serum	0.8 to 3.2 U/L
Bilirubin (total)	Serum	0.1 to 1 mg /dl
Cholesterol (total)	Serum	150-250 mg/dl
Uric acid	Serum	Males—3.5 to 7.5 mg/dl Females—2.5 to 6.6 mg/dl
Calcium	Serum	8.8-10.5 mg/dl
Chloride	Serum	98-108 mEq/L
Sodium	Serum	135-148 mEq/L
Potassium	Serum	3.5-5.5 mmol/l
Phosphorus	Serum	2.5-4.5 mg%
SgPT	Serum	5.40 IU/L
SgOT	Serum	5.40 IU/L
Creatinine	Serum	Males—0.7 to 1.4 mg/dl Females—0.6 to 1.3 mg/dl
Folate	Serum Erythrocyte	5-25 mg/ml 166-640 mg/ml
Glucose	Serum (fasting) Whole blood Post prandial	70-110 mg/dl 60-90 mg/dl less than 140 mg/dl
Iron (total)	Serum	60-150 µg/dl
Iron binding capacity	Serum	250-400 µg/dl
Iron saturation	Serum	20-55%
Phosphatase acid	Serum	0.2 to 1.8 U/L
Alkaline phosphatase	Serum	25-100 U/L
Protein total	Serum	6-7.8 gm/dl
Albumin		3.2-4.5 gm/dl
Globulin		2.3-3.5mg/dl
Triglyceride	Serum	4-150 mg/dl
Urea nitrogen	Serum	8-18 mg/dl
Magnesium	Serum	1.3 to 2.1 mEq/L
Thyroid hormones Thyroxin T ₄ T ₃	Serum (RIA)	4.1 to 11.3 µg/dl 5 ng/dl 0.6 ng/dl

Table 8-8: Comparison of calcium, phosphorus and alkaline phosphatase levels in common disorders of bone and calcium metabolism

Disease	Serum calcium	Serum phosphorus	Serum alkaline phosphatase
Normal	8.8 to 10.5 mg /dl of blood	2 to 5 mg / dl of blood	1 to 4 units/dl of blood in adult (Bodansky)
Rickets	Usually normal except in tetany	Decreased	Increased 20 to 40 X normal
Osteomalacia	Decreased	Decreased	Little if any changes
Hyperparathyroidism	Marked increased	Usually decreased	Increased 2 to 50 X normal
Paget's disease	Usually normal	Usually normal	Occasionally elevated
Osteogenesis imperfecta	Usually normal	Usually normal	Usually normal
Solitary bone cysts	Normal	Normal	Normal
Metastatic osseous neoplasms	May be elevated	Normal	Normal or may be slightly elevated
Tetany	7 mg/dl blood or less	Normal or elevated	Normal

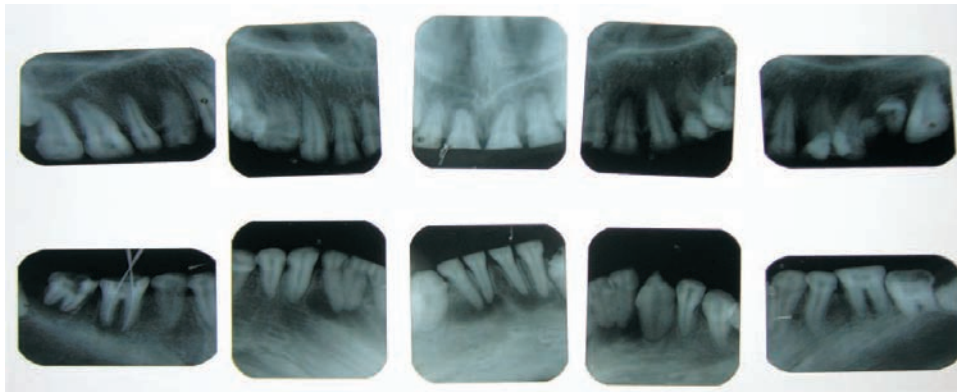


Fig. 8-18: Intraoral periapical radiograph is backbone of dentistry.

currently measured either by protein displacement technique or by radioimmunoassay (RIA). Serum T_4 levels, as measured in this way depend on the availability of thyroxin-binding sites on plasma proteins. When estrogen levels increase, thyroxin binding sites increase in serum and serum T_4 levels increase.

Radiological Investigations

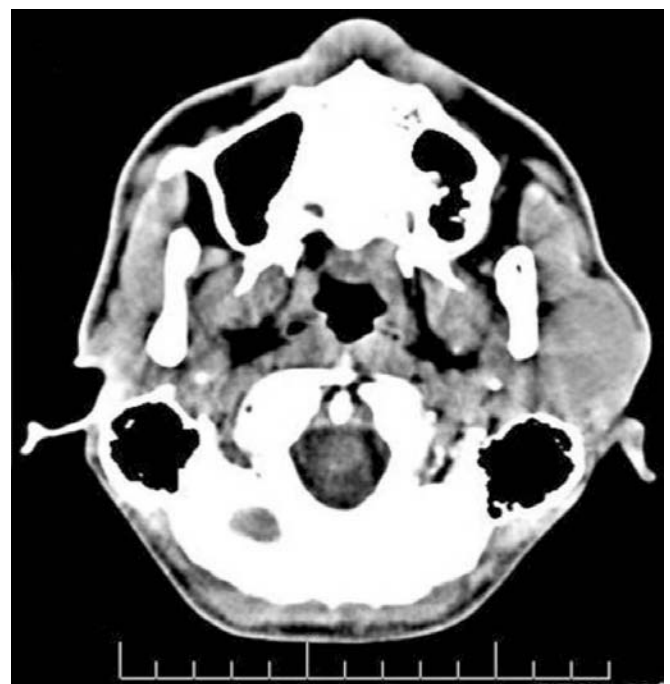
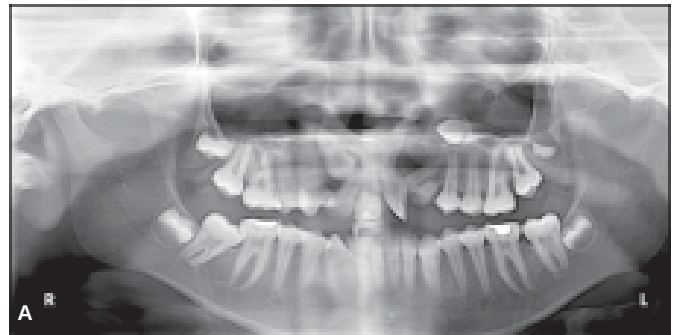
Intraoral Projection

It is the backbone of dental radiography.

- *Intraoral periapical*—it shows all the teeth including surrounding bone and mainly the periapical area (Fig. 8-18).
- *Occlusal*—it shows more areas of teeth and bone than seen in an IOPA radiograph.
- *Bite wing*—it only shows crowns of teeth and adjacent alveolar crests.



Fig. 8-19: Extraoral projection (Water's view) is also indicated in maxillary sinus disorders.



Figs 8-20A and B: Specialized technique like OPG (A) and Computed tomography (B) is also act as valuable aid in making diagnosis of the lesion in oral cavity.

Extraoral Projection

There are many extraoral views which are useful in dental radiography. They are posteroanterior skull, lateral oblique body and ramus of mandible, lateral skull, reverse Towne's, TMJ radiography and Submentovertex view (**Fig. 8-19**).

Special Radiological Examination

Specialized radiographic examination like cephalometric, sialography, OPG, ultrasound, MRI, computed tomography, digital radiography, and radionuclide imaging is carried out (**Fig. 8-20**).

Suggested Reading

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Section **3**

Diseases of Oral Structure



9

Teeth Anomalies

3

Classification

Developmental Tooth Alteration

Size of Teeth

- Microdontia
- Macrodontia

Shape of Teeth

- Gemination
- Twinning
- Fusion
- Concrescence
- Talon cusp
- Dilaceration
- Dens in dente
- Dens evaginatus
- Hypercementosis
- Supernumerary roots
- Paramolar tubercle or Bolk cusp
- Enamel pearl or droplet or nodule
- Globodontia
- Mulberry molar
- Moon's molar
- Hutchison incisor
- Carabelli cusp
- Shovel shaped incisor

Number of Teeth

- Anodontia
- Supernumerary teeth
- Pre-deciduous dentition

Structure of Teeth

- Amelogenesis imperfecta
- Dentinogenesis imperfecta

- Dentin dysplasia
- Regional odontodysplasia
- Dentin hypocalcification

Environmental Tooth Alteration

Environmental Structure Defect

- Enamel hypoplasia

Disturbance of Eruption

- Premature eruption
- Delayed eruption
- Impacted teeth
- Embedded teeth
- Ankylosis of teeth
- Transposition
- Eruption sequestration
- Ectopic eruption
- Premature exfoliation

Postdevelopmental Tooth Loss

- Attrition
- Abrasion
- Erosion
- Internal resorption
- External resorption

Discoloration of Teeth

- Extrinsic stain
- Bacterial stain
- Tobacco stain
- Restorative material
- Medication
- Chlorhexidine stain
- Intrinsic stain
- Erythropoietic porphyria

- Alkaptonuria
- Erythroblastosis fetalis
- Hyperbilirubinemia
- Trauma
- Lepromatous leprosy
- Tetracycline stain

Pulp Disease

- Pulp calcification
- Necrosis of pulp

3

Developmental Disturbances of Teeth

Size of Teeth

Microdontia

It refers to teeth that are smaller than normal. Microdontia is usually transmitted as an autosomal dominant with incomplete penetrance.

Types

- *True generalized*—all the teeth are smaller than normal. It occurs in pituitary dwarfism, Down's syndrome and congenital heart disease.
- *Relative generalized*—normal or slightly smaller than normal teeth; are present in jaws that are somewhat larger than normal. It is hereditary. It often exhibits spacing between the teeth.
- *Localized*—it involves only single tooth. It occurs with congenital heart diseases, Down's syndrome and progeria.

Clinical features

- *Location*—most commonly affected teeth are maxillary lateral incisors and 3rd molars. Supernumerary teeth are frequently smaller than normal.
- *Peg shaped lateral*—it is one of the common form of localized microdontia is peg shaped laterals in which the mesial and distal sides converges or taper incisally, forming peg shaped or cone shaped crown (**Fig. 9-1**).
- *Molars*—when molars are involved, they may also undergo a change in shape from five to four cusps in case of mandibular molar and from four to three cusps in upper molars.

Radiographic features

Radiograph will permit evaluation of size of both, erupted and non-erupted teeth.

Diagnosis

- *Clinical diagnosis*—by looking at the size of tooth clinical diagnosis can be easily made.



Fig. 9-1: Peg shape lateral seen in maxillary region showing cone shaped crown.

Management

- *Crown and bridge work*—crown and bridge work is required for esthetic rehabilitation of teeth.

Macrodonia

It is also called as '*megadontia, megalodontia*'. These are the teeth which are larger than normal.

Types

- *True generalized*—all the teeth are larger than normal. It is commonly associated with pituitary gigantism.
- *Relative generalized*—teeth are normal or slightly larger than normal, but present in a smaller jaw.
- *Localized*—one or more large teeth exist in relation to an otherwise normal dentition and body size.

Causes

- *Facial hemihypertrophy*—it is occasionally seen in facial hemi-hypertrophy, in which half of the teeth in unilateral distribution are affected.
- *Gigantism*—in this generalized overgrowth of the body occurs.
- *Genetic*—genetic component also responsible for macrodonia.

Clinical features

- *Teeth size*—teeth are larger than normal (**Fig. 9-2**).
- *Malocclusion*—there is crowding, which may result in malocclusion.
- *Impaction*—as space is less, there is impaction of teeth.

Radiographic features

The increased size will be demonstrated on the radiograph.

Diagnosis

- *Clinical diagnosis*—by looking at the size of tooth clinical diagnosis can be easily made.



Fig. 9-2: Macrodontia of right central incisor showing large size of tooth.

Management

- *Orthodontic treatment*—if necessary orthodontic treatment is done.
- *Extraction*—if impacted, extraction is indicated.

Shape of Teeth

Gemination

It refers to the process whereby, single tooth germ invaginates resulting in incomplete formation of two teeth that may appear as bifid crown on single root. It occurs during the proliferation stage of the growth cycle of tooth. It has got hereditary and familial tendency.

Clinical features

- *Sex*—males and females are equally affected.
- *Sites*—commonly affected teeth are deciduous mandibular incisors and permanent maxillary incisors.
- *Appearance*—it appears clinically as bifid crown on single root.
- *Number of teeth*—it does not increase or decrease the number of teeth present.
- *Crown features*—there are common pulp canals and either single or partially divided pulp chambers. Crown is wider than normal with shallow groove extending from incisal edge to cervical region. Enamel or dentin of crown of geminated teeth may be hypoplastic or hypocalcified.
- *Complication*—areas of hypoplasia and invagination lines or areas of coronal separation represent caries susceptible area, which may lead to pulpal infection. It may also cause malocclusion and periodontal pathosis.

Radiographic features

- *Cleft crown*—cleft in the crown and invagination are usually outlined by the radiopaque enamel which accentuates them.

- *Pulp chamber*—pulp chamber is single, enlarged and may be partially divided.

Diagnosis

- *Clinical diagnosis*—crown appear as bifid on a single root.
- *Radiological diagnosis*—pulp chamber is single and enlarged

Differential Diagnosis

- *Fusion*—tooth structures with two separate root canals and with either one root or two roots are result of fusion, in contrast to the enlarged and possibly partially divided pulp chamber in an enlarged tooth with bifid crown which is a result of gemination. In gemination the full complement of teeth is present while in the case of fusion, tooth is missing.

Management

- *Reshaping of crown*—affected tooth structure should be removed and crown may be restored and reshaped.
- *Periodic disking*—reduction of mesiodistal width with periodic disking.
- *Crown preparation*—final jacket crown preparation.

Twining

It indicates cleavage of tooth germ which results in formation of supernumerary teeth that is mirror image or near image of tooth from which it has developed.

Fusion

It is also called as 'synodontia'. It represents the embryonic union of normally separated tooth germs. It represents junction at the level of dentin between juxtaposed normal tooth germs. Two separate developing tooth germs being initially close together; as they grow and expand; they contact with each other and the germs fuse to varying degrees.

Etiology

- *Genetic*—it is transmitted as autosomal dominant trait with reduced penetration.
- *Physical*—physical force or pressure generated during development causes contact of tooth germs.

Clinical features

- *Sex*—male to female ratio is 1:1.
- *Location*—it is seen more commonly in anterior teeth. It is more common in deciduous dentition than in permanent dentition. It may occur between a normal tooth and a supernumerary tooth such as 'mesiodens' or 'distomolar'.
- *Size of tooth*—tooth is almost twice in size than normal, with or without bifid crown (**Figs 9-3 and 9-4**).



Fig. 9-3: Fusion of central incisor with lateral incisor in the maxillary region.



Fig. 9-4: Fusion of central and lateral incisor showing large tooth.

- *Root canal*—tooth may have separate or fused root canals.
- *Clinical problems*—it may be related to appearance, spacing and periodontal conditions.
- *Significance*—dental caries is common in fused teeth. It may result in reduced number of teeth in the jaws. When deciduous teeth fuse, the corresponding permanent teeth may be absent.

Radiographic features

- True nature and extent of the union will be more evident on radiograph (**Fig. 9-5**).

Diagnosis

- *Clinical diagnosis*—large tooth with missing adjacent tooth will aid in diagnosis.
- *Radiological diagnosis*—fusion can be seen radiography by dentins.



Fig. 9-5: Fusion of tooth with the dentin seen in incisor area (Courtesy Dr Parate).

Differential Diagnosis

- *Gemination*—it is discussed in Differential Diagnosis of gemination.

Management

- *Endodontic treatment*—morphology of teeth should be determined radiographically for endodontic treatment.
- *Reshaping of tooth*—after endodontic treatment, tooth may be reshaped with a restoration that will mimic independent crown.

Concrescence

It is a form of fusion that occurs after the root and other major parts involved in teeth are formed or when the roots of two or more teeth are united by cementum, below the cemento-enamel junction (**Fig. 9-6**). It is also called as 'false gemination'.

Etiology

- *Developmental*—space restriction during development resulting in extends of cementum deposition between closely approximated roots of teeth.
- *Inflammatory*—in cases of inflammatory damage to the roots of teeth are repaired by cementum.
- *Traumatic injury*—this may also lead to concrescence of teeth.
- *Overcrowding*—overcrowding of the teeth with resorption and interdental bone loss.
- *Distal inclination*—distal inclination of crown of molar will results in contact between cementum resulting in concrescence of the tooth.

Types

- *True concrescence*—if the roots are bound during development.
- *Acquired concrescence*—if the condition occurs after development.

Clinical features

- *Sex*—male to female ratio is 1:1.
- *Site and area*—it is common in maxillary 2nd and 3rd molar area. Either primary or secondary teeth are affected. Usually are involved only two teeth, roots are fused by cementum.
- *Significance*—teeth may fail to erupt or incompletely erupt.
- *Malocclusion and impaction*—there may be malocclusion or the teeth may be impacted.



Fig. 9-6: Concrescence is the fusion of tooth by cementum (Courtesy Dr Alka Kale).

Radiographic features

Diagnosis is made by radiographs. It is not always possible to distinguish between concrescence, teeth in close contact and superimposed teeth (**Fig. 9-7**).

Diagnosis

- *Clinical diagnosis*—it is difficult to differentiate between fusion and concrescence.
- *Radiological diagnosis*—radiograph will show union of two teeth with the help of cementum.

Management

Dentist must be careful while doing extraction.

Talon's Cusp

It projects lingually from cingulum area of maxillary and mandibular teeth or it is an anomalous hyperplasia of cingulum on the lingual surface of maxillary and mandibular incisors, resulting in the formation of supernumerary cusp.



Fig. 9-7: Concrescence showing union of the tooth with the help of cementum (Courtesy Dr Mody).

Pathogenesis

- *Focal proliferation*—a focal proliferation of tissue during development.
- *Exuberant development*—exuberant development of the fourth lobe (cingulum) may occur.

Clinical features

- *Sex*—it may be found in both sexes.
- *Location*—it is common in both dentitions. Most commonly seen on maxillary lateral or central incisor.
- *Appearance*—it resembles like an 'eagle's talon'.
- *Signs*—it blends smoothly with the erupted tooth, except that there is deep developmental groove where the cusp blends with sloping lingual tooth surface.
- *Composition*—it is composed of normal enamel, dentin and contains a form of pulp tissue.
- *Shape*—cusp may or may not contain pulp horn and is usually 'T' shaped (**Figs 9-8 and 9-9**).
- *Significance*—patients can face the problems with esthetic. There is also high incidence of caries. Occlusal interference may be there.
- *Syndrome*—it is associated with Rubinstein-Taybi syndrome and Sturge Weber syndrome.

Radiographic features

- Superimposed with incisors, on which it occurs.
- *Appearance*—outline is smooth and a layer of normal appearing enamel is distinguishable (**Figs 9-10A and B**).

Diagnosis

- *Clinical diagnosis*—T shaped elevation on tooth will easily diagnoses talon cusp.



Fig. 9-8: T-shaped talon cusp seen in maxillary lateral incisor (arrow).



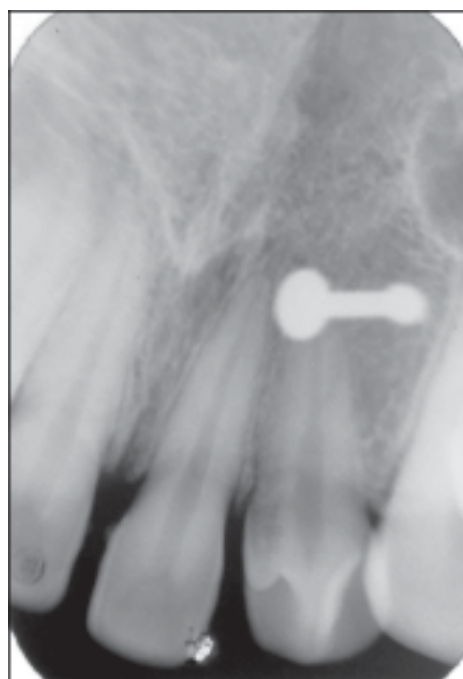
Fig. 9-9: Talon cusp seen as T shaped on the tooth (Courtesy Dr Alka Kale).

Differential Diagnosis

- *Supernumerary teeth*—close association can be determined by tube shift technique.

Management

- *Restoration*—prophylactic restoration of the groove should be done to avoid early carious lesion.
- *Endodontic therapy*—removal of cusp followed by endodontic therapy.
- *Periodic grinding*—periodic grinding should be done to maintain vitality of teeth. After grinding, exposed dentin should be coated with desensitizing agents like fluoride varnish.



A



B

Figs 9-10A and B: Talon cusp is seen as radiopacity with smooth outline

Cusp of Carabelli

It is accessory lingual cusp located on the mesiopalatal cusp of maxillary second deciduous molars and 1st, 2nd and 3rd permanent molars (Fig. 9-11).

It may be unilateral or bilateral, with marked deviation in size. In some cases, accessory cusp is seen occasionally on mandibular permanent or deciduous molar. This is called as *protostylid*.



Fig. 9-11: Cusp of Carabelli seen (arrow) in upper first molar.

Dilaceration

It refers to angulations or sharp bends or curve in the roots and crowns of the teeth.

Etiology

- **Trauma**—mechanical trauma to calcified portion of partially formed teeth results in the displacement of calcified portion in dilaceration. The portion formed after accident is in different direction causing the dilaceration.
- **Development defect**—in some cases, it may be form as developmental anomalies.

Clinical features

- **Sex and site**—it is found equally in both sexes. It is most commonly found in maxillary incisors.
- **Appearance**—curve or bending occurs anywhere along the length of tooth, sometimes at cervical portion or midway along the root or even just at the apex of root (Fig. 9-12).



Fig. 9-12: Unusual bend in the root is seen in case of dilaceration (Courtesy Dr Mody).

- **Significance**—sometimes, angles are so acute that a tooth does not erupt. If the defect is in the crown of an erupted tooth, the angular distortion will be recognized.

Radiographic features

- **Location**—it will show angular distortion of unusual relationship between coronal and radicular portion of the tooth, on either side of defect (Fig. 9-13).
- **Root resorption**—there may be inappropriate resorption of deciduous tooth which will delay the eruption of permanent teeth.
- **Appearance**—if the malformed teeth bend over the fold, the defect will be obscured. If the root bends mesially or distally condition will clearly appear on radiograph.
- **Buccal and lingual dilaceration**—when it is tilted buccally or lingually, dilacerated portion will appear at apical end as a rounded opaque area with dark shadow in central region by apical foramen. Periodontal space about this is evidenced as a radiolucent halo.



Fig. 9-13: Bending of root occur in the middle portion of the teeth.

Diagnosis

- **Clinical diagnosis**—not possible.
- **Radiological diagnosis**—radiograph will show curved root.

Differential Diagnosis

- **Fused root**—in this case, you will be able to find two different root canals.
- **Condensing osteitis**—the tooth is non-vital in case of condensing osteitis.

Management

- **Difficult extraction**—difficulty at the time of extraction.
- **Restoration**—dilacerated crown has to be restored improve esthetics and function and to preclude dental caries and periodontal disease.

Dens in Dente

It is also called as 'dens invaginatus' or 'dilated composite odontome' or 'gestant odontome'. Infolding of the outer surface

of the tooth into its interior surface occurs. It is a developmental variation which is thought to arise as a result of an invagination in the surface of crown before calcification.

Etiopathogenesis

- *Focal growth retardation*—according to Kronfeld, there is relative retardation in growth of a portion of the enamel organ. Due to this, part of the tooth remains stationary and the remaining grows around it causing invagination.
- *Active proliferation*—according to Swanson and McCarthy, it occurs due to proliferation of enamel organ at the inner epithelium apically into dental papilla during the stage of differentiation of developing tooth germ. It grows into dental papilla, as a sort of adenoma.
- *Continuous differentiation*—Rabinowitch said that, it may be formed by continued differentiation of some cells of the inner enamel epithelium.
- *Increased pressure*—study shows that there is presence of extravascular fluid in the soft tissue that fills the potential invagination cavity of the tooth, before it erupts, suggesting that there is increased venous pressure within the invagination. It could be due to pressure on the blood vessels as they pass through the entrance channel of the invagination cavity where enamel is forming concentrically and centripetally, thus tending to progressively narrow the entrance. Expansion of the invagination could then result from the increased venous pressure and transudation.
- *Local causes*—it can occur due to the infection of the deciduous predecessor or trauma. Pressure on the growing teeth can also cause invagination.

Classification

1st classification

- *Coronal dens invaginatus*—it is anomalous infolding of enamel organ into dental papilla. It results in the fold of hard tissue within the tooth, characterized by enamel lining the fold and covering the dentin peripheral to it.
- *Radicular dens invaginatus*—it is a result of invagination of Hertwig's epithelial root sheath resulting in accentuation of normal longitudinal root grooves. It is lined by cementum. Root sheath may bud off sac like invagination that results in a circumscribed cementum defect in root.

2nd classification depending upon site of invagination given by Ohler

- *Type I*—invagination limited to the crown.
- *Type II*—invagination extending to cemento-enamel junction.
- *Type III*—invagination extends beyond the cemento-enamel junctions. It is commonly seen in 2nd premolar either periapical tissue or in the periodontal ligament.

3rd classification given by Aguilo et al

- *Type I*—bifid crown with single root (the crown is larger than normal with notch on the incisal edge and bifid pulp chamber)
- *Type II*—large crown with large root (the crown is larger than normal and there is no groove present. The root is wider than normal with single large pulp canal.
- *Type III*—two fused crown with single root (there are two crown with vertical groove. Root is conical and pulp chamber are separate).
- *Type IV*—two fused crown with two fused root (two crown with vertical groove. The cervical portion are joined along the pulp chamber).

Clinical features

Coronal dens invaginatus

- *Age and sex*—it is seen in children and adolescents. It is more common in females.
- *Site*—commonly affected tooth is permanent maxillary lateral incisor and maxillary central incisor. The lower incisor or cuspid is the next common site. This condition is frequently bilateral.
- *Appearance*—in some cases, there appears to be a grossly magnified cingulum rising to the level of the incisive edge of the tooth, but lacking the normal contour of a cingulum. The labial face of the tooth is often bulbous. Some teeth with these abnormalities are so misshapen as to defy verbal description (**Fig. 9-14**).
- *Crown features*—the crown may or may not be enlarged. The shape of crown may be conical or it may be of irregular shape.
- *Mild form*—there is a deep pit in cingulum.
- *Moderate*—in this type, pocket of enamel is formed within tooth, with dentin at periphery. Opening to the surface



Fig. 9-14: Dens invaginatus showing infolding of outer surface of tooth.

is constricted or remains open. Food debris may become packed in this area with resultant caries and infection of pulp.

- *Severe form*—it may exhibit an invagination extending nearly to the apex of the root.

Radicular dens invaginatus

- *Site*—it is more common in 1st mandibular premolar, upper lateral incisor and second molar. Abnormality is usually unilateral. It occurs most frequently at the site of anatomical defect. It is rare phenomenon.
- *Crown features*—crown is small, short and conical with small orifice at the extreme summit of the convexity. Lingual marginal ridge is prominent.
- *Root features*—cavity is separated from the pulp chamber by a thin wall and opens in oral environment through very narrow constriction.

Radiographic features

Coronal dens invaginatus

- *Radiodensity*—infolding is recognized by its greater radiodensity (**Fig. 9-15**).
- *Size*—it may vary in size from very small and superficial, to large and deep.
- *Pulp chamber*—in addition to small but otherwise normal pulp chamber, there is sometimes an additional cavity of variable shape and size which may be separated from real pulp chamber or may unite with it.

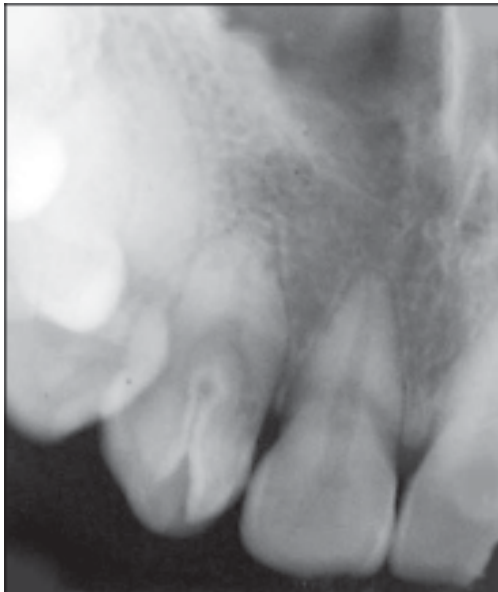


Fig. 9-15: Dens in dente showing infolding of the enamel in the crown of tooth presented as greater radiodensity.

Radicular dens invaginatus

- *Radiodensity*—radicular invagination will appear as a poorly defined, slightly radiolucent structure running longitudinally within the root.

- *Appearance*—the root is composed of a saucer shaped expansion, the concavity of which represents the dilated feature of the tooth.
- *Inverted open umbrella*—the tooth resembles 'inverted open umbrella', the handle of which is short, being represented by the conical, nipple-shaped crown (**Fig. 9-16**).
- *Size*—the diameter of expanded portion of the root, at the extreme apex is as much as 2 cm.
- *Apical foramina*—apical foramina of the root is wide.
- *Apical area*—there is always area of radiolucency present in the bone at the end of the radicular portion as a result of rarefying osteitis, which has followed the passage of organism from the mouth upto the canal in the tooth.

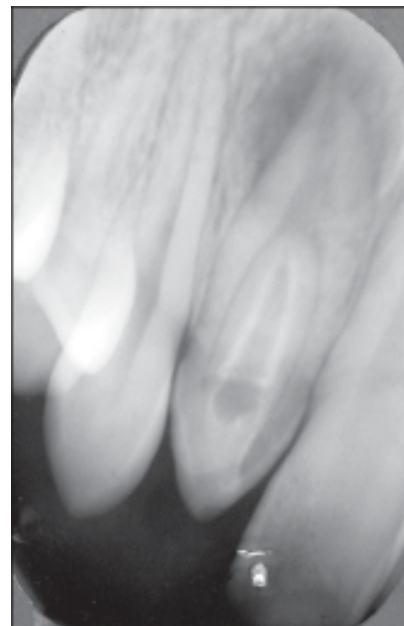


Fig. 9-16: Dens invaginatus showing inverted umbrella appearances (Courtesy Dr Parate).

Diagnosis

- *Clinical diagnosis*—infolding of tooth is seen clinically
- *Radiological diagnosis*—inverted umbrella appearance seen radiographically.

Management

To prevent caries, pulp infection and premature loss of tooth, dense in dente must be treated prophylactically.

Dens Evaginatus

It is also called as 'Leong's premolar', 'evaginated odontome' or 'occlusal enamel pearl'. Dens evaginatus is a developmental condition that appears clinically as an accessory cusp or globules of enamel on occlusal surface, between buccal and lingual cusps of premolar.

Pathogenesis

It is caused by proliferation and evagination of an area of inner enamel epithelium and subsequent odontogenic

mesenchyme into dental organ, during early tooth development.

Clinical features

- **Site**—it occurs on premolar and molar teeth and usually occurs unilaterally or bilaterally.
- **Race**—it develops in persons of Mongoloid ancestry.
- **Composition**—it consists of all three dental tissues, i.e. enamel, dentine and cementum.
- **Appearance**—it appears as a tubercle of enamel on occlusal surface of the affected tooth. Polyp-like protuberance in central groove, on lingual ridge of buccal cusp is seen.
- **Significance**—there may be incomplete eruption. Displacement of teeth with pulp exposure and subsequent infection may present, following occlusal wear or fracture.

Radiographic features

- **Dentin and enamel**—dentin core is covered with opaque enamel.
- **Pulp**—fine pulp horns may be apparent. Pulpal extension is seen in the cusp of the tooth.
- **Tuberculated appearance**—occlusal surface have tuberculated appearance (**Fig. 9-17**).



Fig. 9-17: Tuberculated appearance seen on the occlusal surface of first molar.

Diagnosis

- **Clinical diagnosis**—tubercle on occlusal surface of enamel is the key to diagnosis.
- **Radiological diagnosis**—tuberculated appearance of occlusal surface.

Management

- **Grinding of tubercle**—if tubercle is a cause of occlusal interference, it should be removed and should be followed by indirect pulp capping with calcium hydroxide.
- **Composite reinforcement**—it should be done to protect a cusp from the fracture.

Shovel Shaped Incisors

It is morphologically an anomaly of the crowns of incisor teeth. It is more common in the maxillary teeth.

The shovel shape is manifested by the prominence of the mesial and distal marginal ridges which enclose a central fossa on the lingual surface of incisor teeth. It has frequently a short root (**Fig. 9-18**).

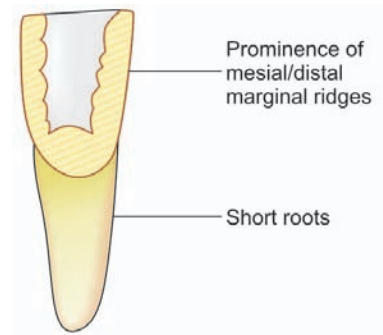


Fig. 9-18: Shovel shaped incisor diagrammatic representation.

Taurodontism

It is described in 1913 by *Sir Arthur Keith*. In this, body of tooth is enlarged at the expense of root. It is characterized by clinical and anatomical crown of normal shape and size, an elongated body and short roots with longitudinally enlarged pulp chambers. Taurodont teeth resembles that of cud-chewing animal (tauro- bull and dont-tooth).

Etiology

- **Hereditary**—it is caused by genetically determined trait.
- **Developmental**—it occurs due to failure of Hertwig's epithelial root sheath to invaginate at proper horizontal level.
- **Mutation**—resulting from odontoblastic deficiency during dentinogenesis of the root.

Classification

- **Hypotaurodont**—it is mild form of Taurodont and it represents slightly apical displacement of pulpal floor.
- **Mesotaurodont**—it is moderate form and pulpal floor displacement is about midline of the root of teeth (**Fig. 9-19**).
- **Hypertaurodont**—it is severe form and apical displacement of pulpal floor is present at the apex.

Clinical features

- **Sex and age**—it is common in early aged men. It has got no sex predilection.
- **Site**—it may affect either deciduous or permanent dentition and teeth involved are invariably molars. It may be unilateral or bilateral, or may exhibit any combination of quadrant involvement.
- **Shape**—involved teeth tend to be of rectangular shape rather than the normal tapering towards root.

- *Associated conditions*—it may associate with certain dermatological condition like epidermolysis bullosa, otodental dysplasia and dyskeratosis congenita.
- *Associated syndromes*—Klinefelter syndrome, Rapp Hodgkin, Down and Trichodontoosseous syndrome.



Fig. 9-19: Moderate form of taurodontism seen in molar teeth (Courtesy Dr Tapasya Karamore).

Radiographic features

- *Pulp and pulp chamber*—pulp chamber is extremely large with much greater apicoocclusal height than normal. Extensions of rectangular pulp chamber occur into elongated body of the tooth. Pulp lacks the usual constriction at the cervix of tooth.
- *Roots*—the root and root canals are exceedingly short. Increased dimension between cemento enamel junction and furcation.

Diagnosis

- *Clinical diagnosis*—rectangular shaped crown.
- *Radiological diagnosis*—extremely large pulp chamber.

Differential diagnosis

- *Developing mandibular molar*—it has got wide apical foramina; the incomplete root and dental papilla will rule out taurodontism.

Management

No specific treatment is necessary as it does not cause any clinical problems.

Supernumerary Roots

Teeth that are normally single rooted exhibit two roots.

Clinical features

- *Sites*—both, maxillary and mandibular molars particularly 3rd molars, exhibit supernumerary roots.
- *Appearance*—they develop as slender outgrowths at the center of furcation area of molar teeth.

Radiographic features

- If the bifurcation produces two distinct apices and these are arranged as one mesial to the other, then it will be seen on the radiographs (**Fig. 9-20**).

- If the two apices are on the labial and lingual side, they may get superimposed on each other appearing as a bulbous root, which may mimic hypercementosis.



Fig. 9-20: Supernumerary root is seen in mandibular first molar.

Differential Diagnosis

- *Hypercementosis*—in a case when root appears bulbous due to superimposition of root on the labial and lingual surfaces, hypercementosis should be ruled out. In a tooth with supernumerary roots, the pulp canal may appear to end a short distance from the blunt square end apex. Arising from blunt end to the canal, are two fine channels which extend one into each root, these channels suggest the presence of two apices, thus excluding hypercementosis.

Management

- Assume significance only during exodontia as these roots may be broken off during extraction.

Paramolar Tubercle or Bolk Cusp

It appears as a nodular elevation. It is seen most frequently on the buccal surface of mesiobuccal cusp in permanent and primary molars.

Enamel Pearls or Droplet, Nodules

It is also called as 'enameloma, ectopic enamel'. It is presence of enamel at unusual location. Pearls or droplets are described as small buttons or nodules of enamel, usually about 1 mm or 2 mm in diameter, that form on the root, or at bifurcation or trifurcation of multirooted teeth.

Pathogenesis

- *Activity of Hertwig's epithelium*—It arises from local activity of remnants of Hertwig's epithelium before it reduces to rests of Malassez.

Clinical features

- *Site*—it is common in trifurcation of maxillary molars, followed by bifurcation of mandibular molars. Those

found on maxillary molars are usually on mesial or distal aspect. In contrast, to those on mandibular molars, which are most often on buccal or lingual aspect.

- **Significance**—rarely, small pulpal extensions may reach into center of nodule and thereby, constitute additional hazard. Rarely, tiny nodule may be sufficiently close to gingival margin to become involved in the periodontal problems.
- **Cervical enamel extension**—they are located on the buccal surface of the root over bifurcation area. In this, localized loss of periodontal attachment with furcation involvement occurs. Cervical enamel extension may also lead to development of buccal bifurcation cysts.

Radiographic features

- **Appearance**—it appears as smooth, round and well defined radiopacity present along the root surface. Radiodensity is same as that of the enamel (**Fig. 9-21**).
- **Pulp**—it may have small core of dentin and rarely pulp horn from the chamber of host teeth.



Fig. 9-21: Enamel pearl seen as rounded opacity on the radiograph.

Diagnosis

- **Clinical diagnosis**—not possible.
- **Radiological diagnosis**—rounded opacity in the furcation area.

Differential Diagnosis

- **Calculus**—clinically detectable.
- **Pulp stone**—can be diagnosed by changing angulation.

Management

- **Maintain oral hygiene**—oral hygiene should be maintained. If it is causing periodontal problems, mass can be removed.

Globodontia or Otodental Syndrome

It is also called as 'otodental dysplasia', 'familial otodontodysplasia'. The term globodontia refers to the enlarged bulbous fused malformed posterior teeth with almost no discernable cusps or grooves. The otodental syndrome also named otodental dysplasia is characterized by a striking dental phenotype known as globodontia, associated with sensorineural high frequency hearing loss and eye coloboma. The condition appears to be inherited in an autosomal dominant mode, although sporadic cases have been reported.

Clinical features

- **Location**—globodontia occurs in both primary and permanent dentition, affecting canine and molar teeth.
- **Age**—age of onset is in fourth decade but it can be seen in early childhood.
- **Appearance of teeth**—Enlarged bulbous malformed posterior teeth with almost no discernable cusps or grooves. Teeth have a round globular or clover leaf appearance (**Fig. 9-22**). The relation between cusps and the major groove is eliminated hence, used the term *globodontia*.
- **Gingiva**—gingiva is inflamed and enlarged. Gingival hyperplasia was a common clinical finding around erupting teeth.
- **Eruption**—there was a significant delay in eruption of the primary and permanent dentition. The primary teeth might exfoliate lately.
- **Decay**—teeth might prone to decay.
- **Malocclusion**—posterior bilateral cross-bite, anterior open bite can also be present.
- **Hearing loss**—there is sensorineural high frequency hearing loss. Hearing loss is progressive and bilateral.
- **Eye defect**—abnormalities ranged from transillumination defects in the inferior iris, due to iris pigment epithelium



Fig. 9-22: Globodontia showing globular pattern with fusion of cusp.

defects, to severe chorioretinal coloboma. Other ocular signs are microcornea, microphthalmos, lens opacity and lens coloboma.

- *Nevi*—several small deeply pigmented lesions are present on faces and scalps.
- *Ears*—ears were protuberant. In some cases ear abscesses are also noted.
- *Stature*—constitutional short stature is also seen.

Radiological features

- Tooth shape is globular with increased radiopacity
- Roots were short and exhibits taurodontism (**Fig. 9-23**)

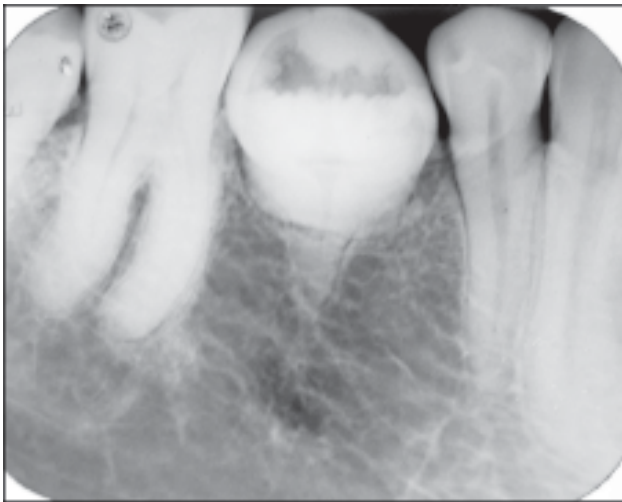


Fig. 9-23: Short roots seen in case of globodontia.

Diagnosis

- *Clinical diagnosis*—globular portion of tooth can be diagnosed clinically.

Management

- Regular follow up, scheduled tooth extraction and eventually orthodontic treatment should be done.

Mulberry Molar

It is a characteristic syphilitic lesion of posterior teeth in which hyperplastic enamel develops with spherical aggregates or globules on the surface of dentin.

Moon's Molar

It is syphilitic lesion of posterior teeth. 1st molar is commonly involved. Cusp of teeth show exaggerated rounded or nodular shapes.

Appearances—it is often grossly distorted, usually narrower mesiodistally with loss of angulation of occlusal edge, which may give cusp a squarish appearance.

Hutchinson's Incisor

Commonly affected teeth are the anterior teeth. Center of the incisal edge shows typical notching. An affected tooth

is screwdriver shaped, with tapering marginal ridges converging towards the incisor edge. Teeth may show barrel shaped outline.

Hypercementosis

It is also called as '*cementum hyperplasia*' or '*exostosis of root*'. It is characterized by deposition of excessive amount of cementum on the root surface. New tissue formation is in direct contact with the cementum of roots of teeth.

Types

- *Localized*—hypercementosis of single tooth. It is usually a reactive, inflammation dependent phenomenon seen on single tooth and usually in relation to periapical osteitis or due to loss of occluding antagonistic tooth.
- *Generalized*—generalized hypercementosis affecting many (all) teeth, but which is seldom recognized as such, occurs with increasing age, i.e. as an age dependent factor. It is also seen as a sign accompanying specific diseases, as for instance, Paget's disease of bone.

Etiology

- *Loss of antagonist*—there is accelerated elongation of tooth occurs due to loss of antagonist. It occurs due to inherent tendency of the periodontium to maintain normal width of the periodontal ligament.
- *Inflammation of the root*—it does not occur at the apex of the root directly adjacent to the area of inflammation, as cementoblasts and their direct precursors in this area are lost. Instead, it occurs at some distance above the apex as the inflammatory reaction acts as stimulation for the cementoblasts.
- *Trauma repair*—occlusal trauma results in mild root resorption and it is then repaired by cementum formation. Root fracture is also repaired on occasion by deposition of cementum between the root fragments as well as in their periphery.
- *Osteitis deformans or Paget's disease of bone*—it is a generalized skeletal disease characterized by excessive amount of cementum formation on roots of teeth and by apparent disappearance of lamina dura of teeth.
- *Others*—other disease which can cause hypercementosis are hyperpituitarism, calcinosis, thyroid goiter, vitamin A deficiency, cleidocranial dysostosis and rheumatic fever.

Clinical features

- *Age*—it is predominately seen in adults.
- *Site*—premolar teeth are often bilaterally affected and symmetrical in distribution. The permanent teeth are affected more commonly than deciduous teeth. In multirooted teeth, one or more roots are involved.

- *Symptoms*—there is no increase or decrease in tooth sensitivity, unless periapical infection is present.
- *Signs*—teeth are vital and not sensitive to percussion. There may be difficulty in extraction of teeth.
- *Fusion of teeth*—in some cases, hypercementosis is so extensive that it causes fusion of two or more adjacent teeth.
- *Roots*—roots appear larger in diameter than normal and present rounded apices.
- *Spike formation of cementum*—it is characterized by occurrence of small spikes or outgrowth of cementum on root surface. It occurs in cases of excessive occlusal trauma probably due to deposition of irregular cementum in focal group of periodontal ligament fibers (Fig. 9-24).

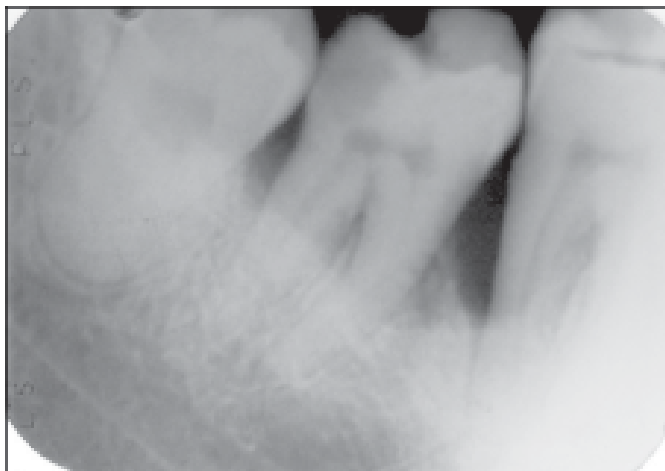


Fig. 9-24: Hypercementosis of third molar seen as excessive cementum deposition around the third molar (Courtesy Dr Ashok L).

Radiographic features

- *Appearance*—there is thickening and apparent blunting of root with rounding of apex. If cementum is distributed eccentrically there is a localized protuberance
- *Apex*—apex appears bulbous in some instances, after symmetrical distribution of cementum.
- *Lamina dura*—lamina dura will follow the outline of teeth in normal periodontal ligament space.
- *Bone resorption*—there may be mildly irregular accumulation of cementum that is accommodated by related area of bone resorption.

Diagnosis

- *Clinical diagnosis*—not significant.
- *Radiological diagnosis*—bulbous appearance of root of tooth due to cementum deposition.

Differential Diagnosis

- *Multirooted teeth and dilacerated root*—successive radiographs from different angle should be taken.
- *Fused root*—recognized by expanded region of root that does not have lower radiodensity of hyperplastic cementum.

Management

- Treatment of the primary cause should be done.

Number of Teeth

Anodontia, Hypodontia and Oligodontia

Several terms are used to discuss number of missing teeth. It is congenital absence of teeth.

Classification

- *True*—it is congenitally absence of teeth
 - *Anodontia*—it is complete lack of tooth development.
 - *Hypodontia*—it is lack of development of one or more teeth (Fig. 9-25).
 - *Oligodontia*—it is lack of development of six or more teeth (Fig. 9-26).
- *False*—it occurs due to extraction of teeth.
- *Pseudo*—it occurs due to multiple unerupted teeth in the jaw.



Fig. 9-25: Hypodontia showing missing teeth in upper anterior region.



Fig. 9-26: Oligodontia showing missing teeth which is more than six in number (Courtesy Dr Parate).

Etiology

- *Genetic*—many hereditary syndromes have been associated with less number of teeth. More commonly they are hereditary ectodermal dysplasia, cleidocranial

dysplasia, craniofacial dysostosis, book syndrome, Coffin-Lowry syndrome, otodental dysplasia etc.

- *Evolutionary trends*—nowadays there is evolutionary trend towards few teeth.
- *X-ray radiation*—this may cause harm to the developing tooth bud resulting in missing teeth.

Clinical features

- *Sex and race*—it is higher in women and most probably in Mongoloid, than whites.
- *Location*—absence may be unilateral or bilateral. Commonly missing are 3rd molar, maxillary lateral incisor (**Fig. 9-27**), maxillary or mandibular 2nd premolar.
- *Associated feature*—associated features include microdontia, reduced alveolar development, increase freeway space and retained primary teeth.



Fig. 9-27: Hypodontia seen in lady patient showing many missing teeth.

Management

- *Orthodontic treatment*—it should be carried out as many patients may develop malocclusion.
- *Prosthesis*—traditional fixed prosthesis and resin bonded bridges should be given.

Supernumerary Teeth

It is also called as '*hyperdontia*'. It is defined as any tooth or tooth substance in the excess of the usual configuration of twenty deciduous or thirty two permanent teeth.

Pathogenesis and etiology

The various factors which are responsible for formation of supernumerary teeth as follows:

- *Phylogenetic reversion theory*—this theory is nowadays discarded.
- *Dichotomy theory*—this theory states that tooth bud is split to form two different tooth,
- *Hyperactivity of dental lamina*—a supernumerary tooth develops from 3rd tooth bud arising from dental lamina near the permanent tooth bud.

- *Hereditary*—it is inherited as an autosomal dominant trait, when associated with syndromes. It is inherited as an autosomal recessive trait when associated with only supernumerary teeth. Supernumerary teeth are most commonly found in relative of affected children than in general population.

Types of supernumerary teeth

It is based on form, and location of teeth.

According to form

- *Supplemental supernumerary teeth*—these teeth duplicate the typical anatomy of posterior and anterior teeth.
- *Rudimentary supernumerary teeth*—these are dysmorphic and can assume *conical* (**Fig. 9-28**) or *tuberculated* forms.
- *Odontome like*—in some cases odontome may be form in place of supernumerary teeth.



Fig. 9-28: Conical type of supernumerary teeth present in anterior maxillary region.

According to location

- *Mesiodens*—it is located at or near the midline in the incisal region of maxilla between central incisors (**Fig. 9-29**). It may occur singly or paired, erupted or impacted or even inverted. It is a small tooth with cone shaped crown and short root. It may cause retarded eruption, displacement or resorption of adjacent root. It frequently causes improper alignment.
- *Distomolar*—it is found in molar region frequently located distal to 3rd molar (**Fig. 9-30**). Generally, these teeth are smaller than normal 2nd and 3rd molar (**Fig. 9-31**), crown morphology is highly abnormal.
- *Paramolar*—it is supernumerary molar, usually small and rudimentary and is situated buccally or lingually to one of the maxillary molars or interproximally between 1st, 2nd and 3rd maxillary molars.
- *Peridens*—supernumerary teeth that erupt ectopically, either buccally or lingually to the normal arch are referred as peridens (**Fig. 9-32**).



Fig. 9-29: Inverted pair of mesiodense present.

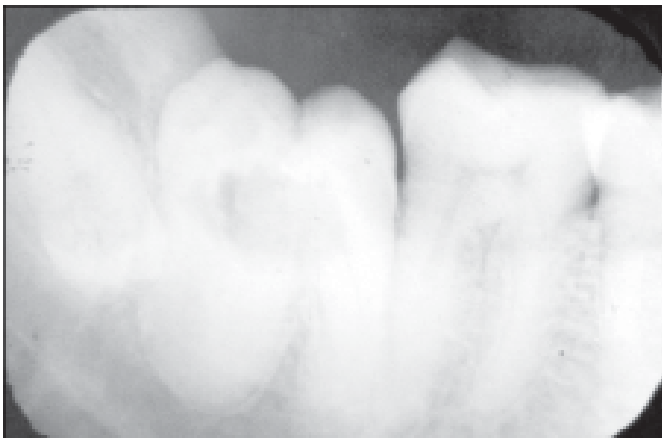


Fig. 9-30: Distomolar seen distal to third molar.



Fig. 9-31: Distomolar seen in maxillary third molar region.



Fig. 9-32: Peridens seen erupted lingually in mandibular premolar region (Courtesy Dr Bhaskar).

Clinical features

- **Prevalence**—the prevalence of supernumerary teeth is about 0.1 to 3.6% of the population.
- **Sex**—males are affected more than females. Male to female ratio is 2:1.
- **Dentition**—it may occur in both dentitions, but frequently found in permanent dentition and more often in mandible.
- **Site**—it is more commonly seen in anterior maxilla (Fig. 9-33A) followed by mandibular premolar region (Fig. 9-33B).



Fig. 9-33A: Supernumerary teeth seen more commonly in maxilla.

- **Effect on dentitions**—supernumerary teeth frequently prevent permanent teeth from erupting. In some cases eruption taken place in abnormal direction. There may be crowding, root resorption of adjacent tooth and malformation of teeth. In some cases supernumerary teeth may be associated with cyst formation.
- **Syndromes**—it is associated with cleidocranial dysplasia, orofacial digital syndrome, Down, Laband, Sturge Weber and Gardner's syndrome.



Fig. 9-33B: Supernumerary teeth seen in mandibular premolar region.

Radiographic features

- *When it is needed*—if abnormal clinical signs are present you can go for OPG examination, IOPA, occlusal radiographic examination.
- *Significance*—radiograph will aid in determining the location and number of unerupted teeth. It can also be used to see if there is any cyst formation.
- *Appearance*—their radiographic picture is characteristic of teeth.

Management

- *Surgical extraction*—it depends on potential effect on normal dentition, their position, number and complications that may result from surgical removal. If required, they should be extracted.

Pre-decious Dentition

It is also called as 'congenital teeth', 'fetal deciduous teeth', 'dentition proceox' and 'natal and neonatal teeth'. There is premature eruption of teeth or teeth like structures that are present at birth.

Natal teeth are the teeth which are present at the time of birth and *neonatal teeth* are the teeth which are present within 30 days after the birth.

Etiology

- *Hereditary*—superficial position of tooth germ.
- *Hormonal influence*—eruption accelerated by febrile incident or hormonal stimulation.

Classification

- *Mature*—they are fully developed in shape and comparable in morphology to the primary teeth. Prognosis is relatively good.
- *Immature*—their structure and development is incomplete. Poor prognosis of teeth.

Clinical features

- *Appearance*—teeth may be conical or may be normal in size and shape and opaque yellow-brownish in color.
- *Signs*—they are hypermobile because of their limited root development. Within relatively short time, premature erupted tooth will become stabilized and other teeth of the arch are erupted. Teeth appear to be attached to a small mass of soft tissue.
- *Significance*—some teeth are so much mobile that there is danger of displacement and possible aspiration and in this case, removal is indicated.
- *Riga fede ulcer*—there is ulceration of the ventral surface of the tongue caused by the sharp incisal edges (Figs 9-34 and 9-35). It leads to interference with proper suckling and feeding and thus the neonate is at risk of nutritional deficiency.
- *Associated syndromes*—it may associate with syndromes like Ellis-van Creveld syndrome and cleft palate.



Fig. 9-34: Ulcer seen on the tongue due to natal teeth.



Fig. 9-35: Neonatal teeth causing ulceration on the ventral surface of the tongue.

Radiographic feature

- Radiographs fail to show any root substance and teeth are very small to represent the normal teeth.

Management

- *Extraction*—extraction of the teeth should be carried out if it is causing inconvenience during suckling, interference with breastfeeding and causing traumatic injury. Extraction should be done after 10 days of life.
- *Rounding of sharp angle*—the other option that may be used is rounding of the sharp angle of incisal edges of teeth.
- *Retaining the tooth*—if not necessary, tooth should not be removed.

Post-permanent Dentition

Persons who have all the permanent teeth extracted and yet have subsequently erupted teeth, particularly after insertion of complete denture come in this category. They possibly develop from bud of dental lamina beyond the permanent tooth germs.

Structure of Teeth**Amelogenesis Imperfecta**

It is also called as 'hereditary enamel dysplasia', 'hereditary brown enamel' and 'hereditary brown opalescent teeth'.

It represents group of hereditary defects of enamel associated with any other generalized defect. It is an ectodermal disease and mesodermal component is normal.

Classification

Classification depends upon the stage of at which the disease occurs. According to it, there are mainly three types of amelogenesis imperfecta

- *Hypoplastic type*—there is defective formation of enamel matrix.
 - *Autosomal dominant*
 - Generalized pitted
 - Localized pitted
 - Diffuse smooth
 - Diffuse rough
 - *Autosomal recessive*
 - Localized pitted
 - Enamel agenesis
 - *X-linked dominant*
- *Hypocalcification type*—there is defective mineralization of formed matrix.
 - *Autosomal dominant*
 - *Autosomal recessive*
- *Hypomaturational type*—in this, enamel crystal lattice remains immature.
 - *Autosomal dominant*
 - *Autosomal recessive*
 - *Snow-capped teeth*.

Clinical features**Hypoplastic type**

It includes localized portions of enamel that do not reach normal thickness during development.

Autosomal dominant

- *Generalized pitted*—it appears as thin enamel on teeth that do not contact with each other mesiodistally. Pinpoint to pinhead pits are randomly distributed over the surface. Enamel on newly erupted teeth is hard with normal yellow-white color. Staining of teeth occurs after exposure to oral environment, giving teeth a black appearance.
- *Localized pitted*—horizontal rows of depressions or one large hypoplastic area with hypocalcification adjacent to and below the hypoplastic area is found. Defects are most prominent on buccal surfaces of the teeth, involving middle 3rd of enamel. Incisal or occlusal surfaces of the teeth are usually not involved.
- *Diffuse smooth*—in this enamel is thin, hard and glossy with smooth surface (**Fig. 9-36**). Enamel is 1/4th to 1/8th of its normal thickness. On newly erupted teeth, they have yellow color, but may vary from opaque white to translucent brown. Some of the enamel may be missing on newly erupted teeth, especially on the incisal and occlusal surface and may be chalky in inter-proximal areas. Delay in eruption occurs with resorption of teeth in the alveolus.



Fig. 9-36: Smooth type of amelogenesis imperfecta showing glossy smooth surface.

- *Diffuse rough*—enamel is hard with rough granular surface that may be chipped from underlying dentin, rather than abrade away as seen with smooth type. Enamel is 1/4th to 1/8th in thickness. Teeth are white to yellow-white when newly erupted. Teeth do not meet at contact points but retain normal tooth outline than the smooth type of imperfecta. It may have thicker enamel at cervical areas. Anterior open bite present.

Autosomal recessive type

- *Localized pitted*—this is severe form than autosomal dominant type. In this, all the teeth are involved in both dentition.
- *Enamel agenesis*—teeth which are erupted have distinct yellow color like normal dentition. Surface is rough and granular, resembling ground glass. There is complete lack of enamel formation. Teeth are widely spaced and do not meet each other at contact point. Patient may have anterior open bite. Numerous teeth are missing in the dentition.

X-linked dominant

- *In males*—there is diffuse, thin smooth shiny enamel in both dentitions. Open contact point. The color varies from brown to yellow-brown.
- *In females*—females will exhibit furrows of thin hypoplastic enamel alternating between bands of normal thickness.

Hypocalcified

In this, enamel matrix is formed normally but without significant mineralization. The enamel is so soft that it can be removed by a prophylaxis instrument.

Autosomal dominant and autosomal recessive

- *Thickness of enamel*—enamel is of normal thickness, although occasional areas of hypoplasia are seen on middle 3rd of labial surface.
- *Consistency of enamel*—the enamel is so soft that it may be lost soon after eruption, leaving crown composed of only dentin (**Fig. 9-37**). Enamel has cheesy consistency and can be scraped from dentin with an instrument or penetrated easily by dental explorer.
- *Color of enamel*—newly erupted teeth are covered with dull lusterless opaque, white, honey colored or yellowish orange or brown enamel.
- *Significance*—exposed dentin may be hypersensitive. Anterior open bite may be present. Patients with this condition are prone to form calculus rapidly.



Fig. 9-37: Hypocalcified type showing loss of enamel in maxillary as well as mandibular teeth (Courtesy Dr Parate).

Hypomaturation type

In this there is defect in the maturation of enamel crystal structure. The enamel can be pierced by an explorer point under firm pressure and can be lost by chipping away from the underlying, normal appearing dentin.

Autosomal dominant

- *Sex*—it is more commonly found in males.
- *Dentition*—both primary and permanent dentitions are affected
- *Color of teeth*—permanent teeth are mottled yellow white in color, but may be darkened with absorption of stains (**Fig. 9-38**). Primary teeth of affected males have ground glass opaque white appearance. Patient occasionally shows slight yellow cast to enamel surface.
- *Intact contact point*—teeth meet at contact points and have normal contour.
- *Thickness of enamel*—enamel approaches normal thickness, but it may be thinner. Point of explorer can be forced into enamel.



Fig. 9-38: Amelogenesis imperfecta of Hypomaturation type showing brown discoloration of teeth.

Autosomal recessive or pigmented type

- *Dentition*—both primary and permanent dentitions are affected.
- *Color of teeth*—enamel has milky to shiny, agar brown color on newly erupted teeth. It may become more deeply stained on contact with exogenous agents.
- *Thickness of enamel*—teeth are of normal thickness and tend to chip away, especially around restoration.
- *Pigmentation*—patient tends to form large amount of calculus which may contain pigment forming agents.
- *Resorption of teeth*—teeth may be seen undergoing resorption within alveolus.

Snow capped teeth

- *Site*—maxillary teeth are affected more commonly than mandibular one.

- *Dentition*—both primary and secondary dentitions are affected.
- *Color of teeth*—in this condition varying amount of enamel on incisal or occlusal aspect of crown is present and has opaque white appearances. Opacity may be solid or flecked and may involve enamel surface. Junctional line of opaque white and translucent enamel is sharp.
- *Dipped into white paints appearance*—pattern of defect on teeth anterior to the posterior teeth resemble that which would be obtained when ‘dipped into white paints’.

Radiographic features

- *Loss of contour*—squarish type of crown being devoid of the normal mesial and distal contours (**Fig. 9-39**).
- *Enamel*—the normal enamel cap is missing and in its place, there is thin and opaque layer of enamel. There may be low and absent cusps, with series of serrations of varying sharpness. There is lack of contrast between enamel and dentin, the whole of the crown presenting a uniform density.
- *Pulp chamber*—if abrasion is advanced then there is obliteration of pulp chamber.
- *Hypoplastic type*—the pitted enamel will appear as areas of mottled density.
- *Hypocalcified type*—in case of hypocalcified type, teeth have moth eaten appearance on radiograph.
- *Hypomaturational type*—in hypomaturational type, enamel is less radiolucent than normal.



Fig. 9-39: Loss of contour seen between the teeth in amelogenesis imperfecta.

Diagnosis

- *Clinical diagnosis*—cheesy consistency of enamel with loss of enamel aid in diagnosis of amelogenesis imperfecta.
- *Radiological diagnosis*—missing enamel cap with low or absent cusp.

Differential Diagnosis

- *Dentinogenesis imperfecta*—Shape and size of crown and root with relatively normal density, obliteration of pulp chamber and root canals in the absence of any marked abrasion should identify dentinogenesis imperfecta.

Management

- *Cosmetic improvement*—it should be done with the help of placement of crown or facial veneer on the teeth.

- *Desensitizing agent*—this should be given to manage the sensitivity which occur to patient due to exposure of dentin.
- *Overdenture*—this should be given to that patient who doesn't have sufficient crown length for the restoration.

Dentinogenesis Imperfecta

There are various names for dentinogenesis imperfecta like ‘hereditary opalescent dentin’ and ‘Capadepon's teeth’. It segregates as an autosomal dominant trait with variable expressivity.

Classification

- *Shield type I*—dentinogenesis imperfecta always occurs with osteogenesis imperfecta. According to Witkop it is called as dentinogenesis imperfecta
- *Shield type II*—It does not occur in association with osteogenesis imperfecta. According to Witkop it is called as ‘hereditary opalescent dentin’.
- *Shield type III*—It has got shell teeth appearance and multiple pulp exposure. According to Witkop it is also called as ‘Brandywine type’.

Clinical features

Shield type I

- *General features*—features of this condition are multiple bone fractures, hyperextensible joints, blue sclera and progressive deafness.
- *Dentition*—deciduous teeth are more severely affected than permanent teeth.
- *Color of teeth*—color of teeth may vary from blue to brownish violet to yellowish brown. Amber translucency of both primary and permanent dentition may be seen (**Fig. 9-40**).
- *Rapid attrition*—enamel may be lost and dentin undergoes rapid attrition (**Fig. 9-41**).



Fig. 9-40: Bluish type of discoloration seen in case of dentinogenesis imperfecta.



Fig. 9-41: Dentinogenesis imperfecta showing rapid loss of enamel (Courtesy Dr Chole).

- **Teeth**—usual scalloping of dentinoenamel junction is absent. The teeth are shorter than normal often markedly, in respect to the roots and crowns. In some cases, there is total crown lost (**Fig. 9-42**).



Fig. 9-42: Total crown lost in the case of dentinogenesis imperfecta (Courtesy Dr Chole).

- **Anterior region**—in the incisor region the crowns tend to more nearly square, but the mesial and distal borders are sometimes curve.
- **Posterior region**—the bicusps and molars are flatter than normal and the normal circumferential curves are accentuated so that the teeth present bulbous appearance. The neck of teeth suddenly narrows down. The appearance of crowns may be described as 'dumpy'.

Shield type II

- Clinical features are same as that of shield type I, except they are somewhat of severe form and it is not associated with osteogenesis imperfecta.

Shield type III

- **Dentition**—both the dentitions are affected.
- **Enamel and dentin**—the thickness of enamel is normal and dentin is very thin.
- **Color and shape**—opalescent color, bell shaped crown and multiple pulp exposure.

Radiographic features

- **Appearance**—constriction of cervical portion of tooth that imparts bullous appearance.
- **Attrition**—there may be slight to marked attrition of teeth.
- **Pulp, root and root canal**—partial or complete obliteration of pulp chamber (**Fig. 9-43**). Pulp obliteration may take place before or after eruption of teeth. Root canals may be absent or thread like or may be blunted.
- **Periapical area**—periapical radiolucency without pulpal involvement and periodontal ligament space widening.
- **Flame shaped pulp canal**—in some cases, the radicular portion of pulp cavities is very narrow, while the pulp chambers have a bulbous expansion terminating in a point deep to the occlusal aspect which resembles 'flame'.
- **Shell teeth**—In Brandywine type, enamel of tooth appears normal, while the dentin is extremely thin and the pulp chambers are enormous. Teeth appear as 'shell teeth'. In addition, root of tooth are extremely short, so it appears as if enamel and dentin surrounded by extremely large pulp chamber and root canals.



Fig. 9-43: Obliteration of pulp chamber seen in dentinogenesis imperfecta (Courtesy Dr Chole).

Diagnosis

- **Clinical diagnosis**—rapid loss of enamel with attrition of tooth. Clinically multiple pulp exposure can be seen.
- **Radiological diagnosis**—obliteration of pulp chamber. Presence of periapical radiolucency without pulpal involvement.

Management

- **Overdenture**—Overdenture should be given and teeth should be covered with fluoride releasing glass ionomer cement.
- **Crown**—cast metal crown in posteriors and jacket crown in anterior can be given.

Dentin Dysplasia

It is a rare disturbance of dentin formation, characterized by normal but atypical dentin formation, with abnormal pulp morphology. It represents as autosomal dominant trait. Some systemic diseases are also manifested as dentin dysplasia features. They are calcinosis cutis, vitaminosis D and tumoral calcinosis.

Classification

According to Shield (clinical)

- Shield type I—dentin dysplasia.
- Shield type II—anomalous dysplasia.

According to Witkop (radiological)

- Radicular dentin dysplasia
- Coronal dentin dysplasia.

Clinical features

Shield type I

- *Synonym*—it is also called as 'rootless teeth', 'non-opalescent and opalescent dentin' and 'radicular dentin dysplasia'.
- *Appearance*—permanent and primary teeth are of normal size, shape and consistency.
- *Color*—affected teeth are occasionally slightly amber and translucent.
- *Mobility*—as the root size is short patient notice mobility of the teeth. There is also premature exfoliation of teeth (Fig. 9-44).
- *Significance*—malalignment and malpositioning due to extreme mobility. Minor trauma may result in exfoliation.



Fig. 9-44: Type I dentin dysplasia showing short root.

Shield type II

- *Color*—significant differences of color in both the dentitions. Primary teeth with yellow, brown, bluish, grey-amber translucent appearances. Permanent teeth of normal color.
- *Root*—root size is normal in size.

Radiographic features

1. Radicular

- *Pulp canals*—pulp chamber and canals are obliterated before eruption and appear as half moon shaped.
- *Roots*—they have short roots with sharp conical and apical constriction. These are abnormally shaped; primary teeth are represented by spicule.
- *Periapical pathology*—multiple periapical radiolucencies are present.
- *Pulp chamber*—obliteration of pulp chamber produce 'crescent or chevron shaped (V shaped line or strip)' pulp remnants.

2. Coronal

- *Thistle tube appearance*—permanent teeth exhibit abnormality, large chamber in coronal portion often described as 'thistle tube' in shape due to radiating extension of pulp chamber.
- *Pulp stone*—teeth show multiple pulp stones.

Diagnosis

- *Clinical diagnosis*—unusual mobility of tooth without periodontal involvement may suspect dentin dysplasia. Radiograph is necessary.
- *Radiological diagnosis*—very short root of the teeth.
- *Laboratory diagnosis*—normal dentinal tubule formation appears to be blocked so that new dentin forms around obstacle and takes the characteristic appearance described as 'lava flowing around boulder'.

Differential Diagnosis

- *Hereditary opalescent dentin*—pulp chamber never fills before eruption in type II dentin dysplasia. Thistle shaped pulp chamber occurs in dentin dysplasia. Teeth in opalescent dentin have bell shaped crown with constriction in cervical region while in dentin dysplasia it is normal. If root is short and narrow it is opalescent dentin, while normal appearing root or no root at all is dentin dysplasia. Periapical rarefaction in association with non-carious tooth is in dentin dysplasia.

Management

- *Oral hygiene*—meticulous oral hygiene should be maintained to avoid early loss of teeth.
- *Retrograde endodontic treatment*—it should be done in patient who have periapical pathology and have a short root.
- *Periapical curettage*—it should be done in cases when there is periapical inflammatory conditions are present.

Regional Odontodysplasia

It is also called as 'odontogenic dysplasia' or 'ghost teeth'. It is an unusual development anomaly in which ectodermal and mesodermal tooth component are affected. It is a

localized arrest in tooth development. In this condition all the elements of tooth are hypocalcified and hypoplastic.

Etiology

It has uncertain etiology. Various factors like trauma, infection, local ischemia, local vascular defect, genetic transmission, local somatic mutation and activation of latent viruses are suggested as etiological agents.

Clinical features

- **Dentition**—deciduous and permanent teeth are involved.
- **Site**—maxillary involved more than mandibular. Most frequently affected are permanent central incisor, lateral incisor and cuspid. Single tooth or several teeth in one quadrant are affected. It may involve both side of midline.
- **Color, shape and size**—affected teeth are small and mottled brown. Shape is irregular with evidence of defective mineralization (**Fig. 9-45**).



Fig. 9-45: Mottled brown discoloration of tooth in regional odontodysplasia.

- **Caries**—affected teeth are susceptible to caries, and prone to fracture.
- **Periapical abscess**—this is frequent finding in regional odontodysplasia. The reason is that discontinuous odontogenic epithelium in the follicle may not completely fuse with oral epithelium which results in invasion of microorganisms in the tooth.
- **Delayed eruption**—there is either delay or total failure of eruption. This occurs as enamel matrix has been shown to induce formation of cementum when it is exposed to extrafollicular tissue. Cementum deposition in crown may retard eruption of teeth.
- **Surface**—surface is rough in feeling
- **Gingival swelling**—gingival swelling adjacent to the tooth surface is common occurrence. The swelling occurs due to calcification in adjacent soft tissue.
- **Associated disease**—this condition may be associated with vascular nevi, hemangioma, hydrocephalus and epidermal nevus syndrome.

Radiographic features

- **Ghost teeth**—there is marked reduction in density, so that the teeth assume ghost appearance. Ghost teeth that do not erupt are so hypomineralized and hypoplastic that they appear to be resorbing.
- **Shell teeth**—sometimes only the shell of enamel remain on the teeth, this appearance is called as shell teeth.
- **Pulp and root**—pulp chamber is very large with wide root canals. Poorly outlined roots are shortened. In some cases, calcification in the pulp is present.

Diagnosis

- **Clinical diagnosis**—irregular shaped teeth with brown discoloration.
- **Radiological diagnosis**—shell teeth appearance seen radiologically.

Differential Diagnosis

Shell teeth of dentinogenesis imperfecta—In dentinogenesis imperfecta, familial involvement in contrast to odontodysplasia. Enamel is not hypoplastic in shell teeth as compared to ghost teeth. Only few teeth are affected in either dentition.

Management

- **Prosthetic replacement**—many suggest to extract involved tooth at the earliest which is followed by prosthetic replacement.
- **Restorative procedure**—in some cases, root canal treatment should be carried out and tooth should be saved.

Environmental Alteration of Tooth

Structure of Teeth

Enamel Hypoplasia

It is an incomplete or defective formation of organic enamel matrix. Local and systemic factors that interfere with the normal matrix formation can cause enamel surface defects and irregularities.

Classification

- **Mild**—there may be only few small grooves, pits and fissures on enamel surface.
- **Moderate**—enamel exhibits rows of deep pits arranged horizontally across the surface.
- **Severe**—considerable portion of enamel may be absent.

Types of environmental hypoplasia

- Hypoplasia due to nutritional deficiency
- Hypoplasia due to exanthematous disease
- Syphilitic hypoplasia
- Hypoplasia due to hypocalcaemia
- Hypoplasia due to birth injury

- Turner hypoplasia
- Dental fluorosis
- Tetracycline hypoplasia.

Hypoplasia due to nutritional deficiency

- *Causes*—it occurs, due to deficiency of vitamin A, C, D, calcium and phosphorus.
- *Age*—2/3rd of this occurs during infancy period or early childhood.
- *Site*—frequently involved are those teeth which are formed within the first year of birth.
- *Pathogenesis*—vitamin D deficiency causes rickettsial phenomenon, resulting from lack of proper calcification of enamel matrix.
- *Appearance*—horizontal pitting occurs in rows, on the teeth undergoing matrix formation at the time of dietary deficiency or during course of febrile episode (**Fig. 9-46**).
- *Color*—pitting characteristically picks up stain and discoloration occurs.



Fig. 9-46: Horizontal pitting seen in anterior region in enamel hypoplasia.

Hypoplasia due to exanthematous disease

- *Causes*—it includes measles, chickenpox and scarlet fever. There is temporary elevation of body temperature.
- *Pathogenesis*—temperature may remain elevated for prolonged period of time and under these circumstances, ameloblasts may be adversely affected.

Syphilitic hypoplasia

- *Site*—it involves maxillary and mandibular permanent incisors and 1st molars. Incisors affected are called as 'Hutchinson incisors' and molar are called as 'mulberry molars' (Moon's molar, Fournier's molar).
- *Hutchinson's incisors*—Upper central incisor is screw driver shaped. Mesial and distal surfaces of crown are tapering and converging towards incisal edge of the tooth, rather than towards cervix. In addition, incisal edge is also notched. The cause behind this is the absence of the central tubercle or calcification center.

- *Mulberry molars*—crown of 1st molar in congenital syphilis is irregular. Enamel of the occlusal surface and occlusal third of the tooth appears to be arranged in agglomerate mass of globule, rather than in well formed cusp. The crown is narrower on occlusal surface, than at the cervical margin.

Hypoplasia due to hypocalcemia

- Tetany induced by decreased level of calcium in the blood, which is as low as 6 to 8 mg per ml.
- As calcium is required for normal tooth formation, there is defective formation of the enamel.
- Enamel hypoplasia, is usually of 'pitting' variety.

Hypoplasia due to birth injury

- *Prenatal*—marked enamel hypoplasia affects incisal 2/3rd of enamel on maxillary primary incisors. It is due to gastrointestinal tract disturbances or metabolic disorders in the fetal life, probably during 2nd and 3rd trimester of pregnancy.
- *Neonatal*—a wide band or line of enamel affects the primary teeth of children associated with premature birth or low birth weight. In traumatic birth, it may affect the process of amelogenesis.

Turner's hypoplasia

- *Turner's tooth*—localized type of hypoplasia is caused by local infection or trauma is called as 'turner's hypoplasia' and that tooth is called as 'turner's tooth'.
- *Pathogenesis*
 - *Local infection*—if deciduous teeth become carious during the period when the crown of succeeding permanent tooth is formed, bacterial infection involving periapical tissues may occur and this may disturb the ameloblastic layer of permanent tooth bud, resulting in hypoplastic crown.
 - *Trauma*—when deciduous teeth have been driven into alveolus and have disturbed the permanent tooth bud and if this permanent tooth bud is still being formed, resulting injury may be manifested as yellowish or brownish stains or pigmentation of enamel, usually on labial surface or as true hypoplastic pitting defect.
- *Site*—most commonly affected teeth are permanent bicuspid as deciduous primary molar are most frequently affected carious tooth in deciduous dentition. Anterior teeth are involved less commonly as their crown is formed before the development of any periapical inflammatory lesion in deciduous anterior region.
- *Appearance*—there may be any degree of hypoplasia, ranging from mild brownish discoloration of enamel to severe pitting and irregularity of the crown. Hypoplastic defect may contain cementum, which may be stained yellowish brown.

Dental fluorosis

Drinking water that contains in excess of 1 PPM (part per million) fluoride can affect the ameloblasts during the tooth formation stage and can cause the clinical entity called as 'mottled enamel'. It is due to disturbance in tooth formation caused by excessive intake of fluoride, during the formative period of dentition.

Pathogenesis

- *Formative stage*—disturbance of ameloblasts during the formative stage of tooth development and higher level of fluorides interfere with the calcification process of matrix.
- *Matrix formation stage*—there is diminished matrix production, change of matrix composition and change in ion transport mechanism.
- *Maturation stage*—in maturation phase, there is retention of amelogenin proteins in the enamel structure leading to formation of hypomineralized enamel.

Classification

- *Questionable changes*—it is characterized by occasional white flecking or spotting of enamel.
- *Mild changes*—it is manifested by white opaque areas involving more of the tooth surface.
- *Moderate and severe changes*—showing pitting and brownish staining of the surface and sometimes even corroded appearance.

Clinical features

- *Dentition*—dental fluorosis in primary dentition is less severe as compared to permanent dentition.
- *Color of teeth*—it frequently becomes stained as unsightly yellow to brown color, which is caused by coloring agents from food, medicine and by disintegration of increase protein contain in the hypomineralized parts of the enamel (**Figs 9-47A and B**).
- *Severe fluorosis*—teeth which are moderately or severely affected may show tendency to wear or even fracture of enamel (**Fig. 9-48**). Sometimes, white patches in enamel may become striated, pitted and mottled.

Clinical classification for fluorosis (TF)

- *Score 0*—the normal translucency of the glossy creamy-white enamel remains after wiping and drying of the surface.
- *Score 1*—thin white opaque lines are seen running across the tooth surface. The lines correspond to the position of perikymata. In some cases 'snow-capping' of cusps may also be seen.
- *Score 2*—the opaque white flecks are more pronounced and frequently merge to form small cloudy areas scattered over the whole surface.
- *Score 3*—merging of the white lines occurs and cloudy areas of opacity occur to spread into many parts of the surface. In between the cloudy area, white lines can also be seen.



Figs 9-47A and B: Mild type fluorosis seen in anterior region.



Fig. 9-48: Severe type of dental fluorosis showing fracture of enamel in anterior region (Courtesy Dr Chole).

- *Score 4*—the entire surface exhibits a marked opacity or appears chalky white.
- *Score 5*—the entire surface is opaque and there are round pits that are less than 2 mm in diameter.

- **Score 6**—the small pits may frequently be seen merging in the opaque enamel to form bands that are less than 2 mm in vertical height.
- **Score 7**—there is loss of the outermost enamel in irregular areas and less than half of surface is involved.
- **Score 8**—the loss of the outermost enamel involves more than half of the enamel. The remaining intact enamel is opaque.
- **Score 9**—the loss of the major part of outer enamel results in change in anatomic shape of the surface. A cervical rim of opaque enamel is often noted.

Radiographic features

- **Mild form**—when there is slight depression or pit on the mesial or distal borders of the crown, it is apparent on a radiograph. When there is depression on lingual or labial surface, it may be gone unnoticed or present as slightly increased darkness.
- **Moderate form**—more extensive lesion appears as a series of rounded dark shadows crossing the tooth in straight lines.
- **Severe form**—very gross deformity produces a crown that is markedly shriveled at its incisal and occlusal surfaces. Such teeth may be presented as small spikes of dental tissue arising from a short stunted base.

Tetracycline hypoplasia

- **Pathogenesis**—it may be incorporated in calcifying enamel matrix by formation of a tetracycline calcium orthophosphate complex.
- **Color of teeth**—after teeth eruption and exposure to sunlight, discoloration may result, ranging from light yellow to brown (**Fig. 9-49**). Varying degree of hypocalcification may also exist.
- **Prevention**—tetracycline should not be administered during pregnancy and until the child become 8 years old.



Fig. 9-49: Hypoplasia of yellow to brown color in upper anterior region due to tetracycline staining.

Management of enamel hypoplasia

- **Restoration**—the hypoplastic teeth are more susceptible to dental caries than the normal teeth. The restoration is usually confined to area of involvement.
- **Crown**—Chrome steel crown is given in case of severe hypoplasia. Severe forms require composite restoration or full ceramic crown.
- **Bleaching with 30% H₂O₂** (hydrogen peroxide)—this technique is enhanced by microabrasion or grinding of the surface layer.
- **Calcium sucrose phosphate gel**—treatment involves cleaning of the affected teeth with pumice and glycerin, rinsing with water and applying 37% phosphoric acid for 1½ or 2 minutes. The treatment is repeated followed by application of 2% sodium fluoride for 4 minute. Finally, a thick layer of 40% calcium sucrose gel is placed on the affected teeth. The patient was instructed not to rinse or eat for 30 minutes.
- **Desensitizing paste**—it should be used to decrease the sensitivity of teeth which may be due to exposed dentin.

Disturbance in Eruption

Premature Eruption

The eruption of teeth may occur before their normal eruption time. In some cases tooth may be present as natal teeth and neonatal teeth.

Etiology

- **Familial pattern**—this can be occurred as familial disease.
- **Endocrine disorders**—secretion of several endocrine organs like thyroid, adrenals and gonads may alter the eruption rate of teeth. It can also be occurred in adrenogenital syndrome.

Clinical features

- **Sites**—usually only one or two teeth erupt early, mostly deciduous and mandibular central incisors.
- **Symptoms**—they are often well formed and normal in all aspects, except they may be mobile.
- **Premature loss of deciduous teeth**—premature loss of deciduous teeth may give rise to premature eruption of permanent teeth.

Delayed Eruption

In some cases eruption may be delayed. It may occur due to various conditions.

Etiology

- **Systemic conditions**—like rickets, cretinism and cleidocranial dysplasia.
- **Local factors**—like fibromatosis gingiva in which dense connective tissue does not permit the eruption of teeth.

Clinical features

- *Partially impacted teeth*—individual permanent teeth are observed to be delayed in eruption. There may be partially impacted permanent teeth.
- *Deviation in eruption path*—there is deviation in the eruption path of teeth.
- *Pseudoanodontia*—patients may suffer from pseudoanodontia.

Management

- *Extraction of teeth and space maintainer*—extract the primary teeth and use space maintainers until the permanent tooth erupts.

Embedded and Impacted Teeth

Embedded teeth are those which are unerupted, usually because of lack of eruptive force.

Impacted teeth are those prevented from erupting by some physical barrier in eruption path.

Etiology

- *Lack of space*—lack of space due to crowding of dental arch and premature loss of deciduous teeth with subsequent partial closure of the spaces they occupied.
- *Rotation*—rotation of tooth bud result in teeth which are aimed at wrong direction because their axis is not parallel to normal eruption path.
- *Systemic disease*—like osteopetrosis, ectodermal dysplasia, cleidocranial dysostosis, rickets and cretinism can be associated with impactions.
- *Cyst and tumors*—in case of overlying cyst and tumors, teeth may fail to erupt in oral cavity.
- *Trauma*—in some cases trauma may cause impaction of tooth.

Clinical features

- *Site*—most commonly affected teeth are maxillary and mandibular third molars and maxillary cuspids (Fig. 9-50), followed by the premolars and supernumerary teeth. Teeth may be impacted distally, mesially, horizontally etc.
- *Significance*
 - Dentigerous cyst may be associated with impacted teeth and may cause of impacted teeth and destruction of bone.
 - Periodontal pocket formation and subsequent infections may occur.
 - Because of location, impacted tooth may cause resorption of roots of adjacent teeth.
 - There may be periodic pain and trismus when infection occurs around the partially impacted teeth.
 - Referred pain from impacted teeth is also been described.

- In some cases, development of pathologic conditions like cyst and tumors can occur.

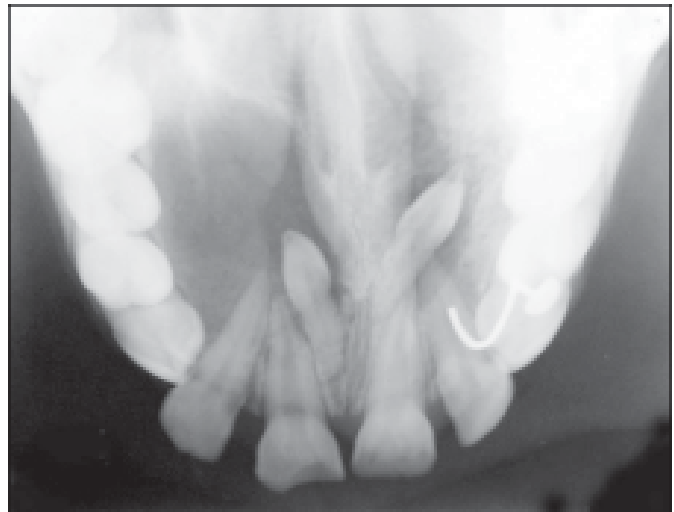


Fig. 9-50: Inverted impacted canine seen in maxillary region.

Impacted mandibular third molar

It may exhibit variety of positions and is classified by Winter as (Fig. 9-51):

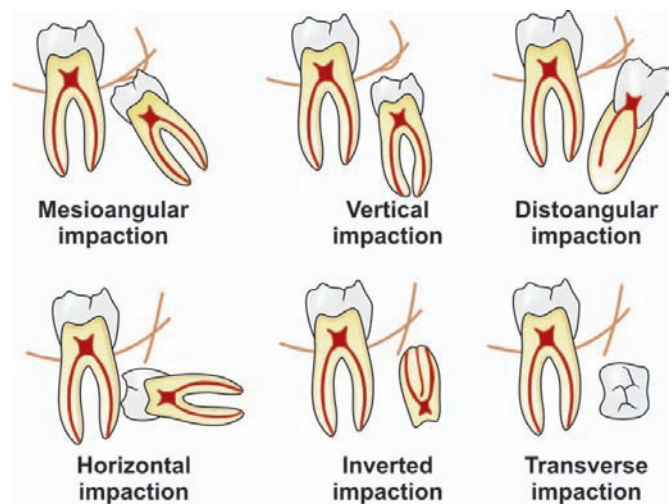


Fig. 9-51: Different types of impaction (diagrammatic representation).

- *Mesioangular impaction*—it lies oblique in bone, the crown pointing in a mesial direction, usually in contact with the distal surface of the root or crown of the second molar (Fig. 9-52).
- *Distoangular impaction*—the third molar lies oblique in the bone, the crown of the tooth pointing distally towards the ramus, the roots are approximate to the distal root of second molars.
- *Vertical impaction*—the third molar is in its normal vertical position but it prevented from erupting by impingement on the distal surface of the second molar or the anterior border of ramus.



Fig. 9-52: Mesioangular impacted of mandibular right and left third molar (Courtesy Dr Parate).

- **Horizontal impaction**—the third molar is in horizontal position with respect to the body of mandible and the crown may, or may not be in contact with the distal surface of the second molar crown or root.

Radiographic assessment of mandibular third molars

- **General**—the main features to be examined include angulations, crown, root, the relationship of the apices with the inferior dental canal, the depth of the tooth in the alveolar bone and buccal and lingual obliquity.
- **Crown**—in crown, you should look for the size, shape, extent of caries and severity of resorption.
- **Root**—in roots, you should note the number, shape, curvature; whether they are favorable or unfavorable and the stage of development.
- **Relationship between canal and root**—the apices of lower 3rd molar often appear close to the inferior dental canal. The roots may be grooved by canal or rarely, tunnel through it (Fig. 9-53).

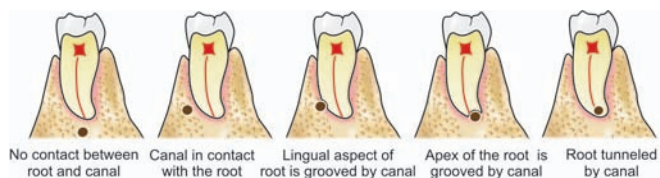


Fig. 9-53: Different types of relationships between canal and roots.

• Tramlines (Fig. 9-54)

- **Normal appearance**—the normal radiographic appearance of canal is too thin radiopaque line called as 'tramlines', which are evident across the root.
- **Loss of tramline**—if there is loss of tramline, then root and canal are in contact.
- **Narrowing of tramline**—if there is narrowing of the tramline, then lingual aspect of root is grooved by the canal.

- **Alteration in direction**—if there is alteration in direction of canal at root apex, then the apex of root is grooved by canal.
- **Radiolucent band in canal**—if there is radiolucent band across the root, the root tunneled through by the canal.

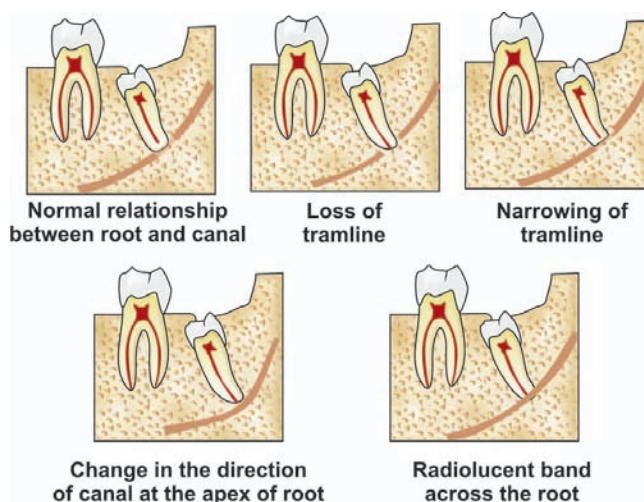


Fig. 9-54: Different types of tramlines appearance seen.

- **Assessment of tooth depth**—two main methods are used commonly to assess the tooth depth i.e. 'winter's line' and using the roots of the second molar as a guide.
- **Winter's line**—in this method, there are three imaginary lines (described by number or color) on a geometrically accurate periapical radiograph (Fig. 9-55).
 - **White line**—the line or white line is drawn along the occlusal surface of the erupted first 1st and 2nd molars.
 - **Amber line**—the second or amber line is drawn along the crest of the interdental bone between the 1st and

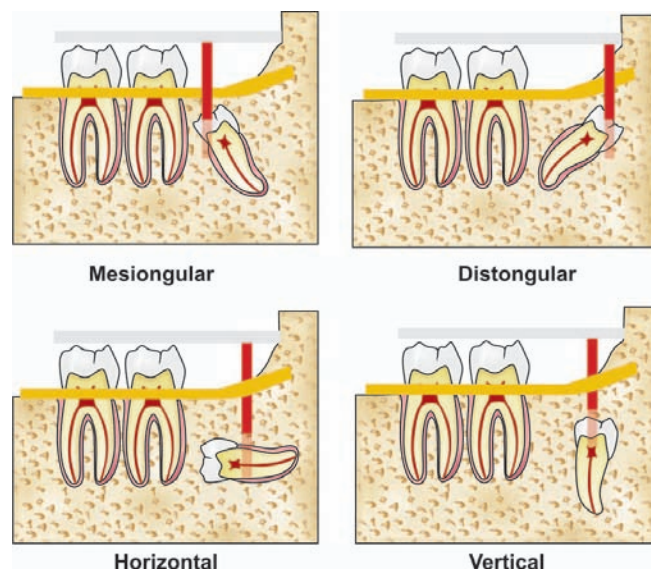


Fig. 9-55: Application of Winter's line to impacted third molar.

2nd molars, extending distally along the internal oblique ridge. This line indicates the margins of the alveolar bone surrounding the tooth.

- **Red line**—the third line or red line is a perpendicular dropped from the white line to the point of application of elevator, but it is measured from the amber line to this point of application of elevator. This line measures the depth of third molar within the mandible. As a general rule, if the red line is 5 mm or more in length, the extraction is considered to be sufficiently difficult.
- **Using the roots of second molars as a guide**—in this, the roots of the adjacent second molar are divided horizontally into three parts. A horizontal line is drawn from the point of application for an elevator to the second molar. If the point of application lies opposite the coronal, middle or apical third, the extraction assesses is classified as being easy, moderate or difficult.
- **Bone**—after this, assessment of surrounding bone is done and checked for the anteroposterior position of the ascending ramus, determine the access to tooth and amount of overlying bone, the texture and density of bone and evidence of previous pericoronal infections.

Management

- **Surgical exposure**—it depends upon the tooth involved. In some cases, like in maxillary cuspids, orthodontic treatment with surgical exposure can be done to bring the tooth in normal occlusion.
- **Surgical removal**—it is done when impacted tooth is causing problems to the patient.
- **Complication**—it includes transient and permanent sensory loss, alveolitis, trismus, infection, fracture, periodontal injury and injury to adjacent teeth.

Ankylosis or Submerged Teeth

Submerged teeth are deciduous teeth that have undergone variable degree of root resorption and then have become ankylosed to bone. Ankylosis of the teeth should be considered as interruption in rhythm of eruption. Ankylosis occurs due to anatomic fusion of cementum or dentin with alveolar bone. Unerupted permanent teeth may become ankylosed by enostosis of enamel.

Etiology

- **Familial pattern**—observations of ankylosed teeth in the same family are very frequent.
- **Changes in local metabolism**—this will lead to ankylosis of tooth in some cases.
- **Trauma**—trauma may cause disruption of periodontal ligament which act as natural barrier between cementum and bone and which prevent osteoblasts from applying bone directly to the cementum.
- **Infection**—some infections may disrupt the normal eruption pathway resulting in ankylosis of tooth.

- **Abnormal tongue pressure**—abnormal tongue pressure may lead to ankylosis of tooth.
- **Other cause**—other causes of ankylosis are chemical and thermal irritation, local failure of bone growth, injury.

Clinical features

- **Age**—it is more commonly found in the second decade of life.
- **Site**—most commonly affected teeth are mandibular deciduous first molars, followed by anterior teeth. Patient who has one or two ankylosed teeth is more likely to have other teeth to be ankylosed.
- **Infra-occlusion**—gradual loss of occlusal plane as the tooth is submerged below the level of occlusion.
- **Delayed eruption of permanent teeth**—exfoliation and subsequent replacement by permanent teeth is prevented due to ankylosis.
- **Lack of mobility**—affected teeth lack mobility even after root resorption.
- **Percussion**—it produces characteristic solid sound in contrast to dull, cushioned sound of normal teeth on percussion.
- **Significance**—development of malocclusion, local periodontal disturbances and dental caries occurs.

Radiographic features

- **Absent periodontal ligament space**—partial absence of periodontal ligament. A break in continuity of periodontal ligament, indicating an area of ankylosis, is usually evident (**Fig. 9-56**).



Fig. 9-56: Ankylosis of third molar showing absence of periodontal ligament space.

Management

- **Surgical excision**—keep tooth under observation. If required, surgical excision is carried out.

Transposition

Tooth may be found occupying an unusual position in relation to other teeth, in the dental arch, i.e. two teeth apparently exchanging their position.

Clinical features

- **Location**—permanent canine is most often involved, with its position interchange with lateral incisor. Second premolar is infrequently found between first and second molar. Transposition of central and lateral incisor is rare. Transposition does not occur in primary dentition (Figs 9-57A and B).



Figs 9-57A and B: Transposition of canine occur with premolar in maxillary left region.

Radiographic features

- It can be recognized on radiograph by the unusual sequence of teeth in dental arch (Fig. 9-58).

Management

These teeth can be prosthetically altered to improve function and esthetics.

Eruption Sequestrum

As the molar teeth erupt through bone, they will occasionally separate small osseous fragments of bone like corkscrew. If bony spicule is large or eruption is fast, complete resorption of bone cannot occur.



Fig. 9-58: Transposition of canine with premolar in maxillary region.

Clinical features

- **Location**—it is a tiny irregular spicule of nonviable bone overlying the crown of an erupting permanent molar. Spicule directly overlies the central occlusal fossa, but is contained within the soft tissues.
- **Signs**—as the tooth continues to erupt, the cusps emerge and the fragments of bone completely sequester through mucosa and are then lost.
- **Symptoms**—child may complain of slight soreness produced by compression of soft tissues over the spicule, by the movement of the spicule in the soft tissue crypt during mastication and following eruption through mucosa.

Radiographic features

- It appears as a tiny irregular opacity overlying the central occlusal fossa, but separated from the tooth itself.

Management

- Removal of spicule, if necessary.

Ectopic Eruption

In this condition, eruption of the teeth does not occur at normal location.

Etiology

- **Large size of teeth**—if the tooth is larger than the normal mean size of all maxillary primary and permanent teeth then chances of ectopic eruption increases.
- **Small jaw size**—smaller maxilla or posterior positioning of maxilla in relation to the cranial base can also lead to ectopic eruption.
- **Local factors**—abnormal angulation and delayed calcification.

Clinical features

- **Age and sex**—it occurs most frequently in boys than girls.

- **Location**—position of teeth can be seen lingually to the normal location (Fig. 9-59).
- **Root resorption**—eruption of permanent 1st molar into roots of primary 2nd molar may cause destruction of distal root of maxillary 2nd molar.
- **Exfoliation**—it may become hopelessly locked and produce premature exfoliation.



Fig. 9-59: Mandibular central incisor seen more lingually than its normal location.

Management

Brass looped wires in contact area should be given.

Premature Exfoliation

Etiology

- **Syndrome**—it is seen in Papillon-Lefèvre syndrome.
- **Hereditary**—it occurs in familial juvenile periodontitis and familial fibrous dysplasia.
- **Nutritional disorders**—like hypophosphatasia and cyclic neutropenia.
- **Others**—histiocytosis and acrodynia.

Clinical features

- **Bone loss**—there is widespread loss of supporting alveolar bone.
- **Teeth**—loosening, migration and spontaneous loss of teeth (Fig. 9-60).

Management

No treatment, except to correct the etiological factors.

Loss of Tooth Structure

Attrition

It is the physiologic wearing away of teeth because of tooth-to-tooth contact, as in mastication. It plays an important physiological role as it helps to maintain an advantageous crown-root ratio and gains intercoronal space of 1 cm, which facilitates third molar eruption. Attrition can be



Fig. 9-60: Early exfoliation seen in deciduous tooth (Courtesy Dr Shetty).

considered pathological when it cause functional, esthetics and dental sensitivity problems.

Types

- **Physiological attrition**—attrition which occurs due to normal aging process, due to mastication.
- **Pathological attrition**—it occurs due to certain abnormalities in occlusion, chewing pattern or due to some structural defects in teeth.

Etiological factors for pathological attrition

- **Abnormal occlusion**
 - **Development**—malocclusion and crowning of teeth, may lead to traumatic contact during chewing, which may lead to more tooth wear.
 - **Acquired**—due to extraction of teeth. Extraction causes increased occlusal load on the remaining teeth, as the chewing force for the individual remains constant.
 - **Premature contact**—in case of edge-to-edge contact, pathological attrition can also occur.
- **Abnormal chewing habits**—parafunctional chewing habit like bruxism and chronic persistent chewing of coarse and abrasive food or other substances like tobacco.
- **Occupation**—in certain occupations, workers are exposed to an atmosphere of abrasive dust and cannot avoid it getting into mouth.
- **Structural defect**—in defects like amelogenesis imperfecta and dentinogenesis imperfecta, hardness of enamel and dentin is reduced and such teeth become more prone to attrition.

Clinical features

- **Sex**—men usually exhibit more severe attrition than women due to greater masticatory forces.
- **Sites**—it may be seen in deciduous as well as permanent dentition. It occurs only on occlusal, incisal and proximal surfaces of teeth. Severe attrition is seldomly seen in primary teeth, as they are not retained for any great period. Palabal cusps of maxillary teeth and buccal cusps of mandibular posterior teeth show most wear.

- *Appearance*—the first clinical manifestation of attrition is the appearance of small polished facet on a cusp tip or ridge and slight flattening of an incisal edge.
- *Physiological attrition*
 - *Onset*—physiological attrition begins with wearing of the incisal edge of an incisor, which is followed by the palatal cusp of maxillary molars and buccal cusp of mandibular molars. It commences at the time of contact or occlusion. Physiological tooth surface loss results in a reduction, in both vertical tooth height and horizontal tooth width (**Fig. 9-61**).



Fig. 9-61: Physiological attrition showing wearing of the occlusal surface of the molar teeth.

- *Contact points*—due to slight mobility of teeth in their socket (which is a manifestation of resiliency of periodontal ligament) similar facets occur at contact points.
- *Color of teeth*—when the dentin gets exposed, it generally becomes discolored i.e. brown in color.
- *Signs*—there is gradual reduction in cusp height and consequent flattening of occlusal inclined plane. There is shortening of the length of dental arch, due to reduction in the mesiodistal diameter of teeth. Secondary dentin deposition occurs.
- *Pathological attrition*
 - *Severe tooth loss*—in pathological attrition severe tooth loss is seen (**Fig. 9-62**).
 - *Dentoalveolar compensation*—if attrition affecting the occlusal surfaces of teeth has occurred, then reduction in occlusal face height (vertical dimension of occlusion) and increase in the freeway space could be anticipated. This may be further complicated by forward posturing of mandible. It is often observed, however, that despite overall tooth surface loss, the freeway space and the resting facial height appear to remain unaltered primarily because of dentoalveolar

compensation. This is important with respect to patient assessment. If restoration of worn teeth is being planned then the extent of dentoalveolar compensation would appear to determine the dentist's strategy; defining the need to carry out measures such as crown lengthening, to ensure the same vertical dimension of occlusion and freeway space.



Fig. 9-62: Pathological attrition showing severe tooth loss in anterior region.

Radiographic features

- *Crown*—smooth wearing of incisal and occlusal surfaces of involved teeth is evident by shortened crown image (**Fig. 9-63**).
- *Pulp*—sclerosis of pulp chamber and canals is seen due to deposition of secondary dentin which narrows the pulp canals.
- *Periodontal ligament*—widening of periodontal ligament space and hypercementosis.
- *Alveolar bone*—some loss of alveolar bone.



Fig. 9-63: Severe attrition of teeth showing reduced crown length.

Management

- *Modifying factors*—treatment of patient depends upon degree of wear relative to the age of patient, etiology, symptoms and patient's desire.
- *Habit breaking appliance*—the provision of one of three different sorts of splints could be considered. A soft bite guard can help in breaking a bruxist habit or simply will protect the teeth during the bruxist habit. A localized occlusal interference splint is designed to break the bruxist habit and can be worn easily during the day. A stabilization splint reduces bruxism by providing an ideal occlusion: it also enables the clinician to locate and record centric relation. In case of bruxism, use of night guards may be effective in reducing attrition.
- *Corrective method*—correction of malocclusion, stoppage of tobacco chewing habit and restriction of diet to non coarse food are useful in avoiding attrition.
- *Management of sensitivity and esthetics*—non-caries loss of tooth tissue may require treatment for sensitivity, esthetics, function and space loss in the vertical dimension.

Abrasion

It is the pathological wearing away of tooth substance through abnormal mechanical process caused by external agents. In case of tooth wear which is accelerated by chewing an abrasive substance between opposing teeth, exhibits a features of attrition and abrasion. This is called as *demastication*.

Etiology

- *Abrasive dentifrices*—use of *abrasive dentifrices* can lead to abrasion of the incisal surface.
- *Habitual*—Habitual pipe smoker may develop abrasion on the incisal edges of lower and upper anterior teeth. In some cases habitual opening of bobby pins may lead to abrasion.
- *Horizontal tooth brushing*—horizontal tooth brushing may lead to abrasion of the cervical area of teeth.
- *Occupational*—it occurs when objects and instrument are habitually held between the teeth by people during working. *Holding nails* or *pins* between teeth e.g. in carpenters, shoemakers or tailors.
- *Dental floss or tooth picks injury*—improper use of dental floss and tooth picks.
- *Ritual abrasion*—it is mainly seen in Africa.

Clinical features

Tooth brush injury

- *Sites*—it usually occurs on exposed surfaces of roots of teeth. It is more commonly seen on left side of right handed persons and vice versa.

- *Mechanism*—it occurs due to back and forth movement of brush with heavy pressure causing bristles to assume wedge shaped arrangement between crown and root.
- *Appearance*—in horizontal brushing there is usually a 'V' shaped or 'wedge' shaped ditch on the root at cemento enamel junction (**Fig. 9-64**). It is limited coronally by enamel.



Fig. 9-64: Wedge shaped ditch seen in the cervical area of anterior teeth.

- *Symptoms*—patient develops sensitivity as dentin becomes exposed.
- *Signs*—the angle formed in the depth of the lesion as well as that of enamel edge is a sharp one. Cervical lesions caused purely by abrasion have sharply defined margins and a smooth, hard surface. The lesion may become more rounded and shallow, if there is an element of erosion present.
- *Dentinal features*—exposed dentin appears highly polished (**Fig. 9-65**). Exposure of dentinal tubules and

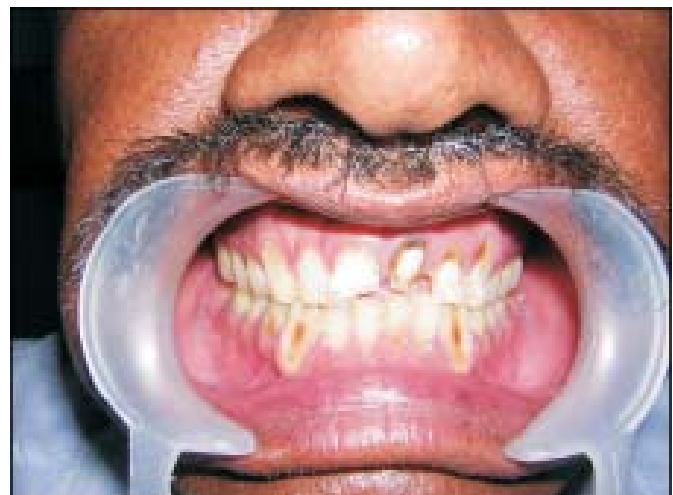


Fig. 9-65: Exposed dentin is seen as highly polished surface.

consequent irritation of the odontoblastic processes stimulates secondary dentin formation which is sufficient to protect the pulp from clinical exposure.

Dental floss or tooth pick injury

- **Site**—Cervical portion of proximal surfaces, just above the gingival margin, is affected. Grooves on distal surface are deeper than on mesial surface (Fig. 9-66).



Fig. 9-66: Abrasion seen on the proximal surface of the patient due to dental floss injury.

Radiographic features

Tooth brush injury

- **Location**—radiolucent defect at the cervical level of teeth.
- **Shape**—well defined semilunar shape, with borders of increasing density (Fig. 9-67).
- **Pulp**—pulp chamber may be partially or fully sclerosed in severely affected teeth.

Dental floss injury

- **Appearance**—narrow semilunar grooves in the interproximal surfaces of teeth near cervical area.



Fig. 9-67: Semilunar defect seen at the cervical area of anterior teeth.

Management

- **Modified teeth cleaning habits**—modification of teeth cleaning habits will be indicated.
- **Removal of cause**—elimination of causative agent should be carried out.
- **Restoration**—restoration should be done for esthetics purpose and to prevent further tooth wear.

Erosion

It is the loss of tooth substance by chemical process that does not involve known bacterial action. Dissolution of mineralized tooth structure occurs due to contact with acids. Erosion is a chemical process in which the tooth surface is removed in the absence of plaque.

Types (depending upon etiology)

- **Intrinsic**—erosion that occur due to intrinsic causes e.g. gastroesophageal reflux, vomiting.
- **Extrinsic**—erosion occurring from extrinsic sources e.g. acidic beverages, citrus fruits.

Etiology

- **Local acidosis**—it is seen in periodontal tissue from damage due to traumatogenic occlusion.
- **Chronic vomiting**—complete loss of enamel on lingual surfaces of teeth through dissolution by gastric hydrochloric acid. Vomiting can also occur in alcoholics, peptic ulcer, gastritis, pregnancy and drug side effect.
- **Acidic foods and beverages**—Large quantities of highly acidic carbonated beverages or lemon juice can produce erosion. Most of the fruits and fruits juices have a low pH and can cause erosion. Frequent consumption of carbonated drinks, which are acidic in nature, may result in the erosion of teeth.
- **Anorexia nervosa**—it induces chronic vomiting often after bouts of uncontrolled eating that is interspersed between periods of starvation, because of inner rejection of food.
- **Occupational**—workers involving in manufacturing of lead batteries, sanitary cleaners or soft drinks can develop erosion.
- **Poorly monitored pH swimming pool**—in cases of poorly monitored pH swimming can also cause erosion of the teeth.
- **Medication**—medication like chewable vitamin C and aspirin tablet may lead to erosion of teeth.

Clinical features

- **Sites**—It occurs most frequently on labial and buccal surfaces of teeth; some times, may occur on proximal surfaces of teeth. Usually confined to gingival thirds of labial surface of anterior teeth. Erosion may involve several teeth of dentition. From extrinsic source, it causes erosion on labial and buccal surface and from intrinsic source, it causes erosion on lingual or palatal source.

- *Appearance*—it is usually a smooth lesion which exhibits no chalkiness.
- *Symptoms*—loss of enamel often causes hypersensitivity in teeth and may also trigger secondary dentin formation.
- *Signs*—loss of tooth substance is manifested by shallow, broad, smooth, highly polished and scooped out depression on enamel surface adjacent to cemento-enamel junction. When erosion affects the palatal surfaces of upper maxillary teeth, there is often a central area of exposed dentine surrounded by a border of unaffected enamel. In most cases, it results in little more than a loss of normal enamel contour, but in severe cases, dentin or pulp may be damaged.
- *Pink spot*—there may be pink spot on tooth which is attributable to the reduced thickness of enamel and dentin making the pink hue of pulp visible.
- *Cupping*—erosive lesions cause 'cupping' in dentin.

Radiographic features

- It appears as radiolucent defect in the crown margins may be well defined or diffuse.

Management

- *Diet control*—in a patient where loss of tooth surface is essentially caused by erosive fluids, advise regarding diet and use of sugar free chewing gum.
- *Fluoride mouthwash*—prescription of a fluoride mouthwash is certainly indicated here.
- *Brushing habits*—brushing habits should be modified.
- *Restoration*—restoration of the defect, usually by glass ionomer cement.
- *Systemic management*—for systemic management of vomiting, patient should be referred to the physician.

Abfraction

It is also called as 'stress lesion'. It is the loss of tooth structure that results from flexure which is caused by occlusal stresses. The magnitude of tooth tissue loss depends on the size, duration, direction, frequency and location of the forces.

Causes and mechanism

- *Eccentric forces*—it has been suggested that the stress lesion or abfraction is a consequence of eccentric forces on the natural dentition. In this case when occlusal force is applied eccentrically on the tooth, it gets concentrated at the cervical margin of the tooth. This will cause flexure which causes stress at the cervical fulcrum and results in loss of the overlying tooth structure. Thus this theory propounds tooth fatigue, flexure and deformation via biomechanical loading of the tooth structure, primarily at the cervical region.

- *Occlusal restoration*—some suggested that occlusal restoration may lead to weakening of tooth ability to resist the stresses of occlusion leading to abfraction.
- *Predisposing factors*—factors, such as erosion and abrasion may play a significant role in tooth tissue loss.

Clinical features

- *Location*—it usually affects buccal/labial cervical areas of teeth. Commonly affects single teeth with excursive interferences or eccentric occlusal loads.
- *Appearance*—it appears as deep, narrow V-shaped notch. The lesion is typically wedge shaped with sharp line angles, but occlusal abfraction may present as circular invaginations.

Resorption of Teeth

It is a chronic progressive damage or loss of tooth structure due to the action of cells called odontoclasts. It can be *physiological* as in case of root resorption of deciduous teeth or *pathological*, which occurs in permanent teeth. Pathological resorption may be *external* or *internal*. In external resorption odontoclasts are located in periodontal ligament and in case of internal resorption cells are located in the dental pulp.

External Resorption

It is lytic process occurring in the cementum or cementum and dentin of the roots of teeth.

Types

- *External resorption*—resorption occurring at the apex or along the lateral surface of the roots.
- *External internal resorption*—in this external resorption is very severe and it leads to involvement of pulp.

Etiology

- *Periapical infection*—periapical granuloma (arising due to pulpal infection or trauma), causes subsequent resorption of root apex. On the radiograph, it appears as slight raggedness or blunting of the root apex in the early stages.
- *Reimplanted teeth*—It may result in severe resorption of root. Tooth root is resorbed and replaced by bone which produces ankylosis. Many implanted teeth exhibit complex resorption of root and are gradually exfoliated.
- *Tumors and cysts*—resorption due to tumors and cysts appears to be essentially by pressure phenomenon. In most cases, tissue is present between the tumor and the tooth and it is from this tissue, cells chiefly osteoclasts arise and initiated root resorption. Cysts like apical periodontal cyst may exert such pressure on the apex of tooth the intervening connective tissue may in turn get stimulated for osteoclasts formation and thus, resorption

begins. The apex may be lost leaving a flat or scalloped surface at the distal end of the root.

- *Excessive mechanical and occlusal forces*—usually, due to the force that is applied during orthodontic treatment, the patient exhibits multiple areas of root resorption, irrespective of the manner of treatment. Pressure from occlusal forces results in destruction, primarily of bone, then small lacunae often appear on the surface of cementum and ultimately, it extends into the dentin.
- *Impacted teeth*—teeth which are completely impacted or embedded in the bone, occasionally, will undergo resorption of the crown and root. Impacted tooth may cause resorption of roots of adjacent teeth without itself getting resorbed. This is commonly seen in case of horizontally or mesioangularly impacted mandibular 3rd molars impinging on roots of 2nd molar.
- *Overhanged root canal filling material*—in such cases not only the apex get resorbed but more or less, sides of the root are affected. In some cases, all or most of the roots disappear and the regenerating bone eventually embraces the root canal filling intimately, so that it appears as if the crown of the tooth is held in position by the root filling.
- *Idiopathic*—in this case, burrowing type of external resorption can occur which is usually seen in relation with single or multiple erupted teeth.
- *Other causes*—other factors which may cause external resorption are dental trauma, grafting of alveolar clefts, hormonal imbalance, intracoronary bleaching of pulpless teeth, Paget's disease and periodontal treatment.

Clinical features

- *Sites*—the most frequent site for external resorption is upper incisors, upper and lower bicusps.
- *Symptoms*—the affected tooth is usually asymptomatic.
- *Signs*—When the root is completely resorbed, the tooth may become mobile. If root resorption is followed by ankylosis then the tooth is immobile, in infraocclusion and with high percussion sound.

Radiographic features

External resorption

- *Appearance*—when the lesion begins at the apex, it causes smooth resorption of the root surface. The conical end is removed and replaced by more or less blunt or usually square apex (**Fig. 9-68**).
- *Bone and lamina dura*—bone and lamina dura show normal appearance. It appears as concave and ragged area on the root surface.
- *Lateral external resorption*—if it involves lateral aspect of the tooth, the lesion will be irregular.



Fig. 9-68: External resorption showing blunt apex of molars teeth.

External-internal resorption

- It appears as eccentrically shaped notch with areas of resorption which are uneven and appears like trabeculae. The resorptions extend apically into pulp or coronally under the enamel. This is also called as invasive cervical resorption.

Differential Diagnosis

- *Internal resorption*—in external resorption, the radiograph shows blunting of the apex, a ragged area, a scooped out area on the side of the root. In internal resorption, one can see root canal with well demarcated enlarged ballooning area of resorption. It appears as expansion of pulp chamber and canals. We can also use SLOB technique for the diagnosis of external resorption. In this, external resorption appear to be shift away from the pulp if the angulation moves mesially.
- *Incomplete root formation*—in this, incomplete canal diverges; whereas in external resorption, they usually converge.
- *Short root*—it has conical apex and the root canal is usually not visible at the extreme apex nor is the foramen.
- *Apicectomy*—it reveals evidence of root filling and the history is available.
- *Foreshortening of root*—despite the short root, the canal at the apical end cannot be seen and neither can be the foramen.

Management

- *Removal of cause*—this is most important management of external resorption.
- *Apicoectomy*—it should be carried out to save the tooth.
- *Curettage and filling*—if the area is broad and on lateral surface, curettage and filling of resorbed area should be carried out.

Internal Resorption

It begins certainly in the tooth. It is a condition starting in the pulp, in which the pulp chamber or the root canals or both, of the tooth expand by resorption of the surrounding dentin.

Mechanism

- Precipitating factor → vascular changes in pulp → inflammation and production of granulation tissue → metaplasia of embryonic connective tissue and macrophages → odontoclasts like giant multinucleated cells → resorption of internal wall of pulp.

Etiology

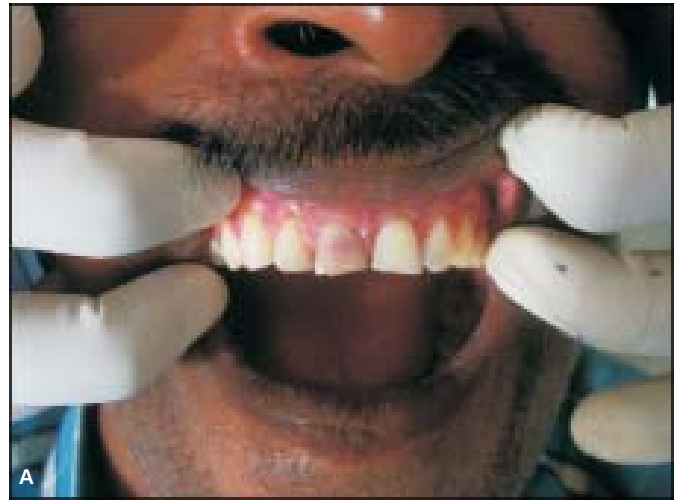
- *Idiopathic*—in this no factors is found to explain the internal resorption.
- *Inflammatory*—inflammatory hyperplasia of the pulp (pulp polyps) may lead to internal resorption of tooth.
- *Pulpal treatment*—pulpal treatment like direct and indirect pulp capping and pulpotomy may stimulate the odontoclasts formation leading to internal resorption.
- *Other causes*—other causes like enamel invagination and acute trauma to teeth may also be responsible for internal resorption.

Types

- *Internal inflammatory resorption*—it occurs due to intense inflammatory reaction within the pulp tissue. In this resorbed dentin is replaced by inflamed granulation tissue.
- *Internal replacement or metaplastic resorption*—it occurs due to absence of any inflammatory reaction within the pulp. In this, pulpal and dentinal walls are resorbed and replaced by bone or cementum like bone.

Clinical features

- *Age and sex*—it occurs during 4th and 5th decades of life and is more common in males.
- *Sites*—it may affect any tooth in primary and secondary dentitions, with prevalence in permanent dentition. It is more common in central incisor, lateral incisors, premolar and canine and 3rd molar according to decreasing frequency. Multiple tooth involvement may be there.
- *Symptoms*—it is asymptomatic and discovered through routine radiographic examination.
- *Pink tooth of mummery*—appearance of pink hued area on the crown of tooth, which represents hyperplastic pulp tissue filling the resorbed area and showing through the remaining overlying tooth substance (**Figs 9-69A and B**). It is called as *pink tooth mummery*.
- *Signs*—when the lesion is located in the crown of teeth, it may expand to such an extent that the crown shows



Figs 9-69A and B: Pink tooth appearance seen in cases of internal resorption of tooth in upper anterior region.

dark shadow due to necrosis of the pulp tissue. If the resorption is in the root, it may weaken the tooth and result in fracture of the tooth.

- *Infectious pulpitis*—it may perforate the crown with hemorrhagic tissue projecting from the perforation and result in infectious pulpitis.

Radiographic features

- *Location*—the destruction may be symmetrically situated around the original canal or it may be eccentric, so that it is situated entirely on one side of the root.
- *Appearance*—radiolucency is homogenous without bony trabeculation or radiopaque foci.
- *Pulp canal*—there is enlargement of canal which may be symmetrical or irregular (**Fig. 9-70**).
- *Shape*—the tooth substance which is destroyed in the root may assume any shape; rounded oval, inverted, pear or irregular (**Fig. 9-71**).
- *Margins*—the margins of enlarged chamber are sharp and clearly defined



Fig. 9-70: Internal resorption in the root showing widening of canal.



Fig. 9-71: Oval-shaped radiolucency in the pulp of internal resorption (Courtesy Dr Mody).

Differential Diagnosis

- *Dental caries*—caries lesion show more diffused margins than internal root resorption. In most cases of internal resorption, enamel is not involved. So, there is no evidence of external orifice.
- *External resorption*—described above

Management

- *Endodontic treatment*—extirpation of pulp with routine endodontic treatment or retrograde filling stops the internal resorption process.
- *Extraction*—extraction of the tooth, if perforation occurs.

Discoloration of Teeth

The color of teeth ranges from yellowish to grayish-white depending on enamel translucency. Discoloration defined

as any change in color or any departure from normal color. Color of teeth becomes darker with age due to increased amount of dentin formation, loss of enamel and staining.

Types

- *Extrinsic*—it occurs due to surface accumulation of exogenous pigment like stains, restorative material, nasmyth membrane, tea, tobacco, iron, medication and chromogenic bacteria.
- *Intrinsic*—it is due to stains within enamel and dentin because of deposition of certain substances. Examples of intrinsic discoloration are amelogenesis imperfecta, dentinogenesis imperfecta, dental fluorosis, porphyria, trauma, medication, ochronosis and hyperbilirubinemia.

Classification

Changes that occur solely in structure and thickness of dental hard tissues:

- *Disturbances of dental hard tissue*
 - Dentinogenesis imperfecta
 - Dental fluorosis
 - Enamel opacities
- Physiologic color changes due to age
- Obliteration of pulp chamber
- Internal resorption
- Initial enamel caries

Discoloration caused by coloring agents taken up by dental hard tissue:

- *During formation*
 - Erythroblastic fetalis
 - Neonatal hepatitis
 - Congenital defect in the bile duct
 - Tetracycline administration
 - Congenital heart disease
- *After eruption*
 - *Endogenous*
 - Necrosis of pulp
 - Hemorrhage of pulp
 - *Exogenous*
 - Amelogenesis imperfecta
 - Turner tooth
 - Dental caries
 - Denudation of dentin
 - Attrition, abrasion and erosion

Various Discoloring Agents

- Food and drink
- Tobacco
- Betel nut
- Restorative materials
- Medicaments like silver, nitrate, tin, iron and iodine
- Blood pigments like hemosiderin, bilirubin and biliverdin
- Products from decomposition of materials.

Causes

Many of the diseases which can cause discoloration of teeth. Some of them are described below.

Obliteration of pulp chamber

Obliteration of pulp chamber may cause by trauma, especially in the formative phase. In this condition, teeth appear *darker* than normal. Maxillary incisors are commonly affected.

Erythroblastic fetalis

It is hemolytic anemia of the newborn caused due to transmission of antibody and excessive hemolysis of erythrocyte.

Erythroblastosis fetalis is based on immunization of Rh-negative mother by Rh-positive red cells of the fetus or perhaps by previous transfusion of Rh-positive blood cells. The mother produces anti-Rh agglutinin. The passage of this soluble substance into the circulation of infant causes complete destruction of the fetal erythrocytes.

Blood pigments like bilirubin and biliverdin are formed which are deposited resulting in stain. Color varies from *green, bluish green to yellow brown or gray*. This is also called as *chlorodontia*. The color of pigment is gradually reduced which is noticed particularly in the anterior teeth. It is transient situation and usually will correct itself with eruption of permanent teeth.

Neonatal hepatitis

Yellowish brown color of primary teeth caused by incorporation of bile pigment during formation of enamel and dentin.

Porphyria

It is an autosomal recessive condition. It is an inborn error in metabolism in which hepatoporphyrin circulating in blood is seen in urine, teeth and bone.

The deciduous and permanent teeth may show *red or brown or pinkish* discoloration (**Fig. 9-72**) which is called as *erythrodontia*. Under ultraviolet light, the teeth always exhibit *red fluorescence*. It occurs due to its physical affinity of calcium phosphate.

Congenital defect in bile duct

In children with this disease, the primary teeth may become discolored by bile pigment, generally *green* discoloration is seen.

Tetracycline Staining

Discoloration of either deciduous or permanent teeth may occur as a result of prophylactic or therapeutic regimen instituted for pregnant females or infants.

Mechanisms

It may cause discoloration during formation period, tetracycline react with calcium to form calcium



Fig. 9-72: Brown discoloration seen in patient of porphyria.

orthophosphate complex. The minimum amount required to produce discoloration is 21 /mg/kg/body weight. It is deposited during mineralization.

Clinical features

It also causes enamel hypoplasia which may be seen in primary as well as in permanent teeth.

It can be demonstrated as *golden fluorescence* in ultraviolet light, which is more intense in dentin than enamel. Bands are more intense toward DEJ. It shows *yellow to brown* discoloration of teeth (**Figs 9-73 and 9-74**). The location coincides with the part of tooth developing at the time of administration of tetracycline.



Fig. 9-73: Tetracycline stain.

It is usually seen with different color in different forms of tetracycline.

- *Chlortetracycline*—gray-brown
- *Oxytetracycline*—yellow
- *Demeclocycline*—yellow
- *Doxycycline*—there is no changes in teeth color.



Fig. 9-74: Yellowish discoloration of teeth due to tetracycline staining.

Congenital Heart Disease

Children with cyanotic heart disease may have maxillary incisors of *milk color* or *bluish violet color*. It may be due to mouth breathing or poorly oxygenated blood.

Oxalosis

It is an autosomal recessive condition. In this disorder, oxalate is deposited in kidney. The *slate gray intrinsic* stain has been demonstrated with odontoblasts, dentinal tubules and within dental pulp.

Discoloring Agents

- *Food and drink*—stain with beverage results in staining on the lingual side of teeth. Foods that contain more chlorophyll produce *green discoloration* of enamel.
- *Tobacco*—tobacco chewing or smoking causes *yellowish brown to black* discoloration of teeth (Fig. 9-75). Tar present



Fig. 9-75: Tobacco stain seen on the lingual surface of teeth.

in the tobacco dissolve in the saliva and penetrates the pits and fissure of enamel.

- *Restorative materials*—incorporation of amalgam in dentinal tubules results in *black to gray discoloration* stains. Deep lingual metallic restoration on anterior incisor can produce grayish discoloration on the labial surface.
- *Medicaments*—silver AgNO_3 used as caries preventing agent and cauterizing agent causes *brownish discoloration* and *black silver* particles in dentinal tubules. Iodine and iron causes *black* discoloration (Fig. 9-76). Copper and nickel can also produce *green* stain.



Fig. 9-76: Black discoloration of the teeth seen due to iron in a female patient.

- *Chromogenic bacteria*—chromogenic bacteria in the oral cavity can stain the teeth to *green, brown, black or orange color*. Bacillus group of bacteria stain the teeth by orange color. Actinomyces species stain the teeth black. Stains from chromogenic bacteria are secondary to the formation of ferric sulfide which is form due to interaction between bacterial hydrogen sulfide and iron in the saliva or gingival crevicular fluid.
- *Chlorhexidine and stannous fluoride stain*—fluoride staining is associated with the use of 8% stannous fluoride. It will produce black stain. Chlorhexidine is associated with yellowish brown stain which involve interproximal surface near the gingival margin.

Different Types of Stains according to Color

Black stain

It usually results due to contact with certain metallic elements such as silver, iron and lead.

It may appear as thin line running approximately 1 mm or so above the gingival margin, it may occur on both the facial and lingual surfaces of teeth.

Stain is firmly attached to the surface but remains extrinsic, thus it may be removed by brush and abrasives. But, it recurs later on.

Green stain

It usually occurs as thick deposit involving the cervical one-third of facial surface of maxillary incisors of young children. It affects boys more frequently than girls.

It is associated with poor oral hygiene and decalcification is sometimes present in enamel, underlying the deposit. It occurs due to chromogenic bacteria or fungi or it may be caused by bacterial action on remnants of enamel cuticle. It is extrinsic and may be removed by simple brushing and abrasive.

Orange stain

It occurs infrequently and usually involve both facial and lingual surfaces of the incisors. It is easily removed than green stain and its cause is unknown but it is believed to be the result of chromogenic bacteria. It is associated with poor oral hygiene and removed with the help of brush and abrasives.

Brown stain (Fig. 9-77)

It can be seen in non-smokers and is usually lighter brown than that of tobacco and form a tenacious, but delicate film on surface of the teeth. It is usually seen more commonly on lingual surface of lower incisors and buccal surface of maxillary molar teeth. It is formed due to altered salivary mucins which have undergone change through the action of bacterial enzymes.



Fig. 9-77: Brown staining occurs in patient due to tobacco consumption and patient is also having fluorosis.

Management**Extrinsic**

Scaling and polishing with paste containing fine powdered pumice.

Intrinsic

Fluoride stains are treated by applying bleaching solution. It consist of

- Anesthetic ether—0.2 ml
- HCl 30%—1 ml etches the enamel surface.
- H₂O₂ 30%—1 ml bleaches enamel.

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Definitions

Congenital

Present at or before birth but not necessarily inherited, i.e. transmitted through the genes.

Hereditary

They are apparent at birth but some may not become evident for years.

The cell consists of cytoplasm and nucleus. Nucleus of each somatic cell contains 23 pairs of chromosome out of which one is the pair of sex chromosome; rest 22 pairs of chromosomes are called as *autosomes*. *Chromosome* is a nuclear structure composed of DNA, which contains units of hereditary gene. *Gene* is a portion of DNA coded for the synthesis of specific protein or polypeptide chain. Gene is determinants of hereditary characteristics. *Locus* is the site occupied by the gene on chromosome. An *allele* is number of alternative form that gene may take.

If both the allele at a given locus is identical to pair, it is called as *homozygous* and if the alleles are different it is called as *heterozygous*. A gene showing trait in heterozygous is considered as *dominant*. Genes are transmitted from one generation to other in a generally predictive way.

Autosomal Dominant Inheritance

- Every affected person has at least one affected parent.
- Males and females are both affected likely.
- There is no skipping of generation.
- Affected person typically transmits trait to their offsprings.

Autosomal Recessive Inheritance

- Appear only in their siblings, not on their offspring parent or other relatives.

- $\frac{1}{4}$ siblings are affected.
- Males and females are equally likely to be affected.

X-linked Dominant

- Since females have twice as many X chromosomes, they exhibit a higher frequency of trait.
- Affected female will transmit the gene to $\frac{1}{2}$ of her offsprings.

X-linked Recessive

- Incidence of the trait is much higher in males than in females.
- Affected man can transmit it to his daughters who are carriers.
- Trait cannot be transmitted from father to son.

Developmental anomalies of the head and neck are originated during transformation of branchial system into adult's derivatives. They are also result from failure of migration of neural crest cells.

Developmental Disturbances of the Jaws

Agnathia

It is also called as '*hypognathous*'. Agnathia, derived from Gnathus meaning the jaws, is total absence of either one or both the jawbones. This is very rare condition.

Clinical Features

- *Absence of mandible*—if mandible is absent, the upper part of the face may be normal and the skin of the lower part will be continuous with the suprasternal integument.

- *Hyoid bone*—the hyoid bone is sometimes absent, despite the presence of a rudimentary tongue.
- *Vertical slit*—in place of buccal orifice, there may be a vertical slit.
- *Deformed ear*—in case of unilateral absence of mandibular ramus, it is not unusual for ear to be deformed or absent.
- *Associated abnormalities*—there may be absence of ears, hypoplastic tongue, cleft palate, dysplastic ears, hypertelorism, microstomia, narrow auditory canal with palpebral fissures slanting down.

Management

- *Surgical reconstruction*—surgical reconstruction though it is difficult it should be carried out.

Micrognathia

Micrognathia is severely deficient jaw, which most commonly affects the mandible. It is common to see individuals having a very small mandible.

Types

- *Apparent micrognathia*- this is not due to abnormality of small jaw, in terms of size but rather to an abnormal positioning or abnormal relation of one jaw to another, which produces illusion of micrognathia.
- *True micrognathia*—It is due to small jaw. It is again classified as
 - Congenital
 - Acquired

Etiology

Congenital

- *Congenital abnormalities*—in many instances it is associated with other *congenital abnormalities*, particularly congenital heart disease and Pierre Robin syndrome (cleft palate, micrognathia, glossoptosis).
- *Forceps delivery trauma*—one of the main causes of this condition is the use of forceps during birth where the baby is pulled out by clamping a pair of forceps on either side of the head. If the joint in this area, called the temporomandibular joint is badly bruised, the mandible does not develop. Sometimes this condition is also associated with congenital heart disease.

Acquired

- *Ankylosis*—acquired type is post-natal type and result from disturbances in the area of *T.M joint* (ankylosis) (Fig. 10-1).
- *Mouth breathing*—mouth breathing can be a predisposing factor for maxillary micrognathia.

- *Agenesis of condyle*—agenesis of condyle also results in true mandibular micrognathia.
- *Posterior positioning*—posterior positioning of mandible with regard to skull or to a steep mandibular angle result in an apparent retrusion of the jaw.



Fig. 10-1: Acquired micrognathia occurs due to ankylosis of jaw.

Clinical Features

- *Micrognathia of maxilla*—it is due to deficiency of premaxillary area and patient with this deformity appears to have the middle third of face retracted.
- *True mandibular micrognathia*—it is uncommon and patient appears clinically to have severe retrusion of chin, steep mandibular angle and deficient chin button.
- *Signs*—micrognathia is one of the causes of abnormal alignment of teeth. This can be seen by observing the occlusion of teeth. Often, there will not be enough room for the teeth to grow. If the upper jaw is short, then, occlusion may be abnormal.
- *Symptoms*—in true micrognathia, the jaw is small enough to interfere with feeding of the infant and may require special nipples in order to feed adequately. There may be difficulty in respiration. Due to the small size of the arch, the jaw is not able to accommodate the tongue, which is forced back into the oropharynx, blocking the air passage.
- *Syndromes associated with micrognathia*—Pierre Robin syndrome, Hallerman-Streiff syndrome, trisomy 13, trisomy 18, Turner's syndrome, Treacher-Collins syndrome and Marfan's syndrome.

Management

- *Orthognathic surgery*—this is recommended treatment modality for micrognathia. This surgery is followed by orthodontic appliance to correct malocclusion.

- *Small maxilla*—if upper jaw is short, then it can be corrected with surgical orthodontic treatment by properly aligning the teeth and then moving surgically and elongating the short maxilla, in order of three to four millimeters of the upper central incisors to show when an individual is smiling.
- *Small mandible*—small mandible can be corrected, depending on the degree of deformity and problem, advancement of mandible and chin surgically.

Macrogathia

It refers to the condition of abnormally large jaws. It is also called as '*megathia*'. Macrogathia literally means a large jaw. The mandible is most often affected in this case giving rise to a condition where the bone protrudes.

Etiology

- *Pituitary gigantism*—there is generalized increase in size of entire skeleton.
- *Paget's disease of bone*—overgrowth of cranium and maxilla occurs.
- *Acromegaly*—progressive enlargement of mandible owing to hyperpituitarism in adults.

Clinical Features

- *Prognathism*—mandibular protrusion or prognathism is common occurrence, which is due to disparity in the size of maxilla to mandible and posterior positioning of maxilla in relation to the cranium.
- *Mandible*—mandible is measurably larger than normal. Increased mandibular body length.
- *Gummy smile*—in certain patients with congenital abnormalities, there may be elongation of maxilla. There is much "show" when the patient smiles, so that there is a so-called "gummy" smile. This is due to the upper jaw being too long.
- *Ramus*—large ramus which forms less steep angle with body of mandible.
- *Chin*—there is prominent chin button.

Management

- *Osteotomy*—resection of portion of mandible to decrease the length, followed by orthodontic treatment.

Facial Hemihypertrophy

It is also called as '*Friedreich's disease*', '*hemihyperplasia*'. It may involve entire half of the body, one or both limbs, face, head and associated structures. It represents the hyperplasia of the tissue rather than hypertrophy of the tissue.

Etiology

- *Endocrine dysfunction*—hormonal imbalance may lead to hemihyperplasia of the face.
- *Genetic*—incomplete twinning, chromosomal abnormalities and localized alteration of intrauterine development also lead to facial hemihypertrophy.
- *Vascular abnormalities*—lymphatic and vascular abnormalities may act as causative factors.
- *Neurogenic*—some neurogenic malformation may act as causative factors for this disease.

Terminology

- *Complex hemihyperplasia*—in this, one whole side of the body is affected.
- *Simple hemihyperplasia*—hyperplasia is limited only to one limb.
- *Hemifacial hyperplasia*—if the enlargement is confined to one side of the face.

Clinical Features

- *Age and sex distribution*—it has vague onset, usually in childhood, adolescence or early adult life. Females are affected more than males.
- *Sites*—it involves the eyelids, cheeks, lips, facial bones, tongue, ears and tonsils. It is more common on right side of body.
- *Symptoms*—occasionally, poorly localized, vague, painful sensation in muscles affected.
- *Appearance*—enlargement of one half of the head present since birth. Enlarged side grows at a rate proportional to uninvolved side (**Fig. 10-2**).
- *Associated abnormalities*—it is associated with other abnormalities like mental deficiency, skin abnormalities, compensatory scoliosis, and hemi-megalocephaly.



Fig. 10-2: Enlargement of one half of face on left side occurs in facial hemihypertrophy.

(hypertrophy of one cerebral hemisphere with ipsilateral ventricular dilatation).

- *Skin lesion*—pigmentation, hypertrichosis, telangiectases and hemangioma may occur on skin.
- *Syndrome associated*—syndromes associated with facial hemihypertrophy are Proteus syndrome and Klippel-Trenaunay-Weber syndrome.

Oral Manifestations

- *Dentition is abnormal in three respects*—crown size, and root may be large. Rate of development of permanent teeth on the affected side is more rapid and erupt before their counterparts on the uninvolved side. Primary teeth shed early.
- *Bone*—bone of maxilla or mandible is also enlarged.
- *Tongue*—tongue is commonly involved and bizarre patterns of enlargement of papilla, in addition to general unilateral enlargement of tongue.
- *Buccal mucosa*—buccal mucosa frequently appears velvety and may hang in soft pendulous folds on affected side.

Radiographic Features

- *Bone enlargement*—enlargement of bone on affected area. The malar bone, zygomatic process and temporal bone may be enlarged in all diameters, in some cases.
- *Alveolar process*—the alveolar process is enlarged in some cases on affected side (**Fig. 10-3**).
- *Mandibular canal*—it also increases on the affected side.
- *Teeth*—crown size of the tooth is enlarged.



Fig. 10-3: Enlargement of bone and teeth on the right side of patient (Courtesy Dr Datarkar).

Diagnosis

- *Clinical diagnosis*—enlargement of one side of face can be noticed clinically.
- *Radiological diagnosis*—it will also show enlargement of bone and teeth on one side.

Management

- *Cosmetic repair*—cosmetic surgery should be performed which includes soft tissue debulking, face lifts and Orthognathic surgery.

Facial Hemiatrophy

It is also called as 'Parry-Romberg syndrome', 'Romberg hemifacial atrophy', 'hemifacial microstomia' and 'progressive facial hemiatrophy'. Parry-Romberg syndrome is a rare disorder characterized by slowly progressive wasting (atrophy) of the soft tissues of half of the face (hemifacial atrophy). Wasting is associated with skin, cartilage, connective tissue, muscle and bone. It is described by Parry in 1825 and well documented by Romberg in 1846.

Etiopathogenesis

- *Idiopathic*—In most cases, Parry-Romberg syndrome appears to occur randomly for unknown reasons (sporadically).
- *Familial*—certain studies state that it has got familial occurrence.
- *Localized scleroderma*—in scleroderma there is constriction of tissue which may lead to progressive hemifacial atrophy.
- *Malfunction of sympathetic nervous system*—atrophic malfunction of cervical sympathetic nervous system can lead to neurotrophic change which in turn leads to hemiatrophy.
- *Trigeminal neuralgia*—peripheral trigeminal neuralgia is associated with hemiatrophy in some cases.
- *Loss of adipose tissue*—this will lead to shrinkage of soft tissue.

Clinical Features

- *Age*—onset noted in 1st or 2nd decades of life which are apparently normal at birth. In rare cases, the disorder is apparent at birth.
- *Location*—in most cases, progressive tissue wasting is limited to one half of the face, usually the left side.
- *Onset*—onset is marked by white line furrow or mark on one side of face or eyebrow near midline.
- *Progress*—in most affected individuals, hemifacial atrophy typically progresses over approximately three to five years and then ceases.
- *Coup de sabre*—some patients may show sharp line of demarcation resembling large linear scar between normal and abnormal skin. This is called as coup de sabre.
- *Skeletal like appearance*—affected areas may demonstrate shrinkage and atrophy of tissues beneath the skin

(subcutaneous tissue), in the layer of fat under the skin (subcutaneous fat) and in underlying cartilage, muscle and bone. If disease is not limited then patient may get skeletal-like appearance.

- **Skin**—in addition, the skin overlying the affected areas may become darkly pigmented (hyperpigmentation), in some cases, certain areas of white (depigmented) patches (vitiligo). There may be hollowing of cheek and eyes may appear depressed in the orbits.
- **Neurological manifestation**—this may include severe headache that lasts for extended periods of time and may be accompanied by visual abnormalities, nausea and vomiting (migraine). There are also facial pain due to trigeminal neuralgia. There are periods of uncontrolled electrical disturbances in brain (*seizures*) that usually are characterized by rapid spasms of a muscle group that spread to adjacent muscles (*contralateral Jacksonian epilepsy*).
- **Hair**—there is graying (blanching) of hair as well as abnormal bald patches on the scalp with loss of eyelashes and the middle (median) portion of eyebrows (alopecia).
- **Skull changes**—there is underdevelopment of the base of skull, in some cases the face is affected. When the malar bone is small, the side of the face is flat but an absent malar bone produces a depression inferior to the orbit.
- **Face**—the soft tissue of face are small and thinner than normal.
- **Ear**—aplasia or hypoplasia of external ear. The ear canal is missing.

Oral Manifestations

- **Lip and tongue**—many individuals also experience atrophy of half of the upper lip and tongue.
- **Teeth**—delayed eruption or wasting of the roots of certain teeth on the affected side. This will lead to malocclusion of teeth.
- **Deviation of jaws**—while opening the mouth, jaw is deviated on the affected side.
- **Jaws**—growth of jaws and teeth is affected. Ramus is deficient vertically and there is delay in mandibular angle development. The mandibular body can be shorter than normal.
- **Posterior open bite**—due to mandibular hypoplasia on affected side there may be unilateral posterior open bite.

Radiographic Features

- **Size reduction**—reduction in size of bone on affected side (**Fig. 10-4**). There is also reduction in size of condyle, coronoid process or overall dimension of body and ramus of mandible. The affected side of the face is smaller in all dimensions than the opposite side.



Fig. 10-4: Hemifacial atrophy presenting as reduced size of the jaw on affected side (arrow).

Diagnosis

- **Clinical diagnosis**—white line or furrow on one side and patient is having ‘coup de sabre’.
- **Radiological diagnosis**—reduction of size of bone on affected side.

Differential Diagnosis

- **Mandibulofacial dysostosis**—hereditary pattern is present and cleft palate is also seen.
- **Hemifacial microstomia**—these are congenital and non-progressive conditions.
- **Post-traumatic atrophy**—history is important.
- **Partial lipodystrophy**—it is usually bilateral.
- **Goldenhar syndrome**—it is congenital and non-progressive.

Management

- **Surgical reconstruction**—it primarily involves augmentation of the affected areas. Surgery is deferred unless diseased process is burned out.
- **Orthodontic treatment**—it is given for the correction of malocclusion.
- **Other treatment modalities**—it includes injection of silicones and bovine collagen, inorganic implants and autologous tissue transfer.

Hemi-maxillofacial Dysplasia

It is ‘*segmental odontomaxillary dysplasia*’. Etiology of this condition is unknown and many times, it is mistaken as craniofacial fibrous dysplasia or hemifacial hyperplasia. This condition remains stable.

Clinical Features

- **Age**—it is frequently encountered in younger patient.
- **Symptoms**—there is painless unilateral enlargement of the maxillary bone.
- **Gingiva**—fibrous hyperplasia of gingiva occurs in majority of patient.

- **Teeth**—maxillary premolar are missing and primary teeth in the affected area show enamel defect.
- **Becker's nevus**—some patients may be associated with hypertrichosis and hyperpigmentation (Becker's nevus).

Radiological Features

- **Trabeculae**—trabeculae are thickened and are vertically oriented resulting in granular appearance.
- **Maxillary sinus**—it is smaller in size on the affected side.

Diagnosis

- **Clinical diagnosis**—unilateral enlargement of maxillary bone with fibrous hyperplasia of gingiva will give clue to diagnosis
- **Radiological diagnosis**—sinus is smaller on radiograph.

Management

- **Orthodontic therapy**—some patients may require orthodontic therapy.
- **Orthognathic surgery**—it is rarely recommended in patient with segmental odontomaxillary dysplasia.

Cleidocranial Dysplasia

It is also called as 'cleidocranial dysostosis', 'Marie and Sainton disease', 'craniocleido-dysostosis'.

Etiology

- **Hereditary**—it appears as a true dominant Mendelian characteristic.
- **Genetic**—incomplete penetration of genetic trait. There is defect in CBFA I gene chromosome.

Clinical Features

- **Sex**—the disease affects men and women with equal frequency.
- **Sites**—it primarily affects skull, clavicle and dentition.
- **Shoulder meet in midline**—there may be complete absence of clavicle and patients have unusual mobility. They can bring their shoulders forward until they meet in midline (**Figs 10-5A and B**).
- **Head**—the head is brachycephalic (reduced anterior-posterior dimension but increased skull width) or wide and short.
- **Skull**—in skull the fontanels often remain open or at least exhibit delayed closing and for this reason, tend to be rather large. Open skull suture and multiple wormian bones are present and there is occasional stunting of long bone.
- **Lacrimal and zygomatic bone**—lacrimal and zygomatic bone are also underdeveloped.

- **Sagittal suture**—sagittal suture is characteristically sunken, giving skull flat appearance.
- **Nose**—nasal bridge is depressed with a broad base.



Figs 10-5 A and B: Patient can bring his shoulders forward so that they can meet in midline. Also note the depressed nasal bridge.

Oral Manifestations

- **Micrognathia**—maxilla and paranasal sinus are underdeveloped, resulting in maxillary micrognathia.
- **Maxilla**—maxilla is underdeveloped and is smaller than normal, in relation to mandible.
- **Prognathism**—maxilla is small and mandible is usually normal in size, which gives the appearance of prognathism.
- **Teeth**—prolonged retention of primary dentition and delayed eruption of permanent dentition. Numerous unerupted teeth are found which are most prevalent in the mandibular, premolar and incisor area (**Fig. 10-6**).
- **Palate**—high narrow arched palate and cleft palate may be common.
- **Hypoplasia of enamel**—the crown may be pitted as a result of enamel hypoplasia.



Fig. 10-6: Numerous unerupted teeth seen in patient with cleidocranial dysplasia.

Radiographic Features

- *Skull finding*—skull film reveals open sutures (**Fig. 10-7**), presence of wormian bones, widened cranium, delayed ossification of fontanelles, frontal and occipital bossing and basilar invagination.



Fig. 10-7: Open suture seen in cleidocranial dysplasia patient.

- *Chest examination*—examination of chest reveals malformation and absence of clavicles (**Fig. 10-8**).

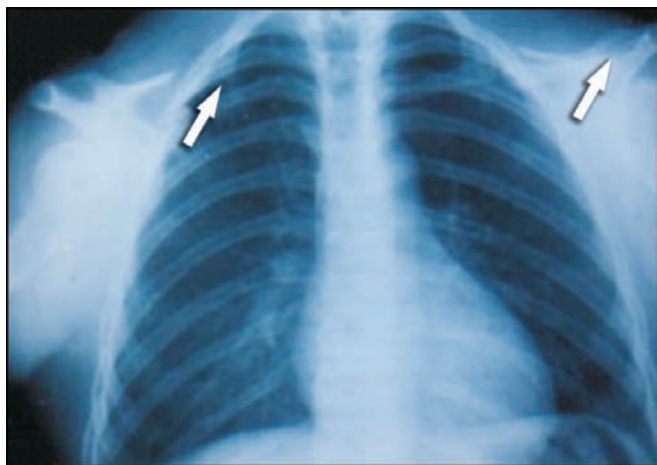


Fig. 10-8: Absence of clavicle seen on chest examination in cleidocranial dysplasia.

- *Teeth*—jaw examination reveals prolonged retention of primary dentition. The unerupted teeth are frequently displaced to occupy an oblique or horizontal position in the jaw.
- *Supernumerary teeth*—presence of supernumerary teeth usually in anterior region (**Fig. 10-9**). The supernumerary teeth in this area often show anomalous and distorted roots.
- *Jaws*—there is small underdeveloped jaw. There is also flattened mandibular angle (**Fig. 10-10**) and overgrowth

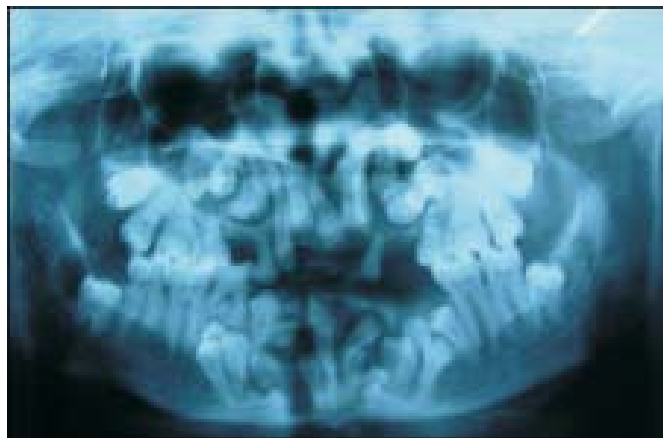


Fig. 10-9: Multiple supernumerary teeth found in cleidocranial dysplasia.

of cranial base; anteroposteriorly. Pointed coronoid process.

- *Roots*—roots of teeth are often some what short and thinner than the normal.



Fig. 10-10: Flattened mandibular angle seen in cleidocranial dysplasia.

Diagnosis

- *Clinical diagnosis*—meeting shoulder in the midline is typical feature of cleidocranial dysostosis.
- *Radiological diagnosis*—supernumerary teeth, open sutures and absence of clavicle are present in cleidocranial dysostosis.

Management

- *Management of skull and clavicle anomalies*—there is no treatment require for this anomalies. Patient can live its normal life.

- **Dental care**—as patient may be having supernumerary teeth, they should be extracted. Prosthetic replacement of teeth should be carried out. Unerupted teeth should be extruded orthodontically.

Craniofacial Dysostosis

It is also called as '*Crouzon's disease or syndrome*'. In some instances, Crouzon syndrome is inherited as an autosomal dominant trait. In other cases, affected individuals have no family history of disease. The disorder is characterized by distinctive malformations of the skull and facial (craniofacial) region.

Clinical Features

- **Age**—Crouzon's syndrome is a rare genetic disorder that may be evident at birth (congenital) or during infancy.
- **Craniosynostosis**—in most infants with Crouzon syndrome, the fibrous joints between certain bones of the skull (cranial sutures) close prematurely (craniosynostosis).
- **Proptosis**—in addition, facial abnormalities typically include unusual bulging or protrusion of the eyeballs (proptosis) due to shallow eye cavities (**Fig. 10-11**)
- **Strabismus**—outward deviation of one of the eyes (divergent strabismus or exotropia); widely spaced eyes (ocular hypertelorism). This may lead to blindness of the patient.
- **Cranial malformation**—the premature closing of suture may result in brachycephaly (short head), scaphocephaly (boat shaped head), or trigonocephaly (triangle shaped head). In extreme cases patient may demonstrate '*clover leaf skull*' (kleeblatt schadel deformity).



Fig. 10-11: Ocular proptosis with triangular frontal defect seen in patient of Crouzon's syndrome.

- **Frontal bone**—bulging of frontal bone in midline, over the nose and downward sloping of back of head. There is protuberant frontal region with an anterior-posterior ridge overhanging the frontal eminence and often passing to the root of nose (triangular frontal defect).

Oral Manifestations

- **Maxillary hypoplasia**—maxillary hypoplasia with shortened anteroposterior dimensions of maxillary arch.
- **Parrot beak**—in some cases, facial angle is exaggerated and the patient nose is prominent and pointed, resembling 'parrot beak' (**Fig. 10-12**)



Fig. 10-12: Parrot beak appearance of the patient seen in Crouzon's syndrome. There is also midfacial hypoplasia.

- **High arched palate**—dental arch width is reduced and this gives an appearance of high arch palate (**Fig. 10-13**).



Fig. 10-13: High arch palate seen in Crouzon's syndrome.

- **Teeth**—unilateral or bilateral cross-bite is evident with open bite and crowding in mandibular teeth. Shovel shaped maxillary incisors cleft lip and palate are also evident (**Fig. 10-14**)

- *Sinus*—the antrum are small and underdeveloped. The mandible is large as compared to maxilla, so there is prognathism.



Fig. 10-14: Unilateral cross-bite on left side in Crouzon's syndrome.

Radiographic Features

- *Absence of sutures*—digital marking in skull as a result of increased intracranial pressure from early synostosis of cranial sutures.
- *Beaten silver appearance*—the cranial walls are thin with multiple radiolucencies appearing as depressions or scalloped appearance of beaten silver (Fig. 10-15).
- *Others*—malformation of calvarium, flattened mandibular angle, conical teeth and partial anodontia.

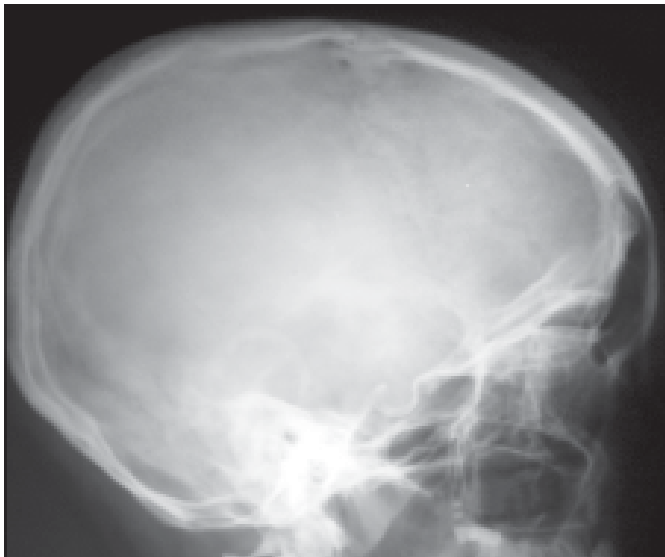


Fig. 10-15: Beaten silver appearance seen in Crouzon's syndrome.

Diagnosis

- *Clinical diagnosis*—high arch palate, with midface deformity, proptosis is a typical feature of Crouzon syndrome.
- *Radiological diagnosis*—beaten silver appearance seen in Crouzon syndrome.

Management

- *Midfacial advancement*—this is done to correct midfacial hypoplasia.
- *Craniectomy*—it is needed to alleviate the raised intracranial pressure.
- *Fronto-orbital advancement*—this is done to correct ocular defect.

Mandibulofacial Dysostosis

It is also called as 'Treacher Collins syndrome' and 'Franceschetti syndrome'. It is often inherited as autosomal dominant trait. It is derived from first and second branchial arch. It results from failure or incomplete migration of neural crest cells to the facial region.

Clinical Features

- *Zygomatic bone*—underdevelopment of zygomatic bone, resulting in midfacial deformities.
- *Eyes*—there is downward inclination of palpebral fissure. There is deficiency of eyelashes. In some cases, eyes assume corresponding slant. A notching (*coloboma*) from the outer third of lower eyelids, There is varying degree of visual impairment in some cases.
- *Ears*—affected infants may also have underdeveloped (hypoplastic) and/or malformed (dysplastic) ears (Fig. 10-16) (pinnae) with blind ending or absent external ear canals (microtia), resulting in hearing impairment (conductive hearing loss). Absence of external auditory canal resulting in partial or complete deafness is also found.



Fig. 10-16: Treacher Collins syndrome showing malformed ear.

- *Malar bones*—the normal prominence of cheek is either missing or reduced depending upon the presence or absence of malar bone. There is usually hypoplasia of malar bone.
- *Neurological disorders*—secondary mental deficiency.
- *Fistulae*—blind fistulae between the angle of ears and the angle of mouth.
- *Associated features*—anal atresia/stenosis, congenital cardiac anomaly, rectovaginal fistula and tracheo-esophageal fistula.

Oral Manifestations

- *Appearance*—underdevelopment of mandible with steep mandibular angle. Due to this, lower anterior teeth stand away from upper teeth, when the mouth is closed.
- *Fish or bird appearance*—facial appearance sometimes resembles fish or bird (**Fig. 10-17**).
- *Micrognathia*—an incompletely developed, abnormally small lower jaw.
- *Macrostomia*—an unusually large mouth.
- *Palate*—there is presence of high arch palate with cleft palate.
- *Malocclusion*—abnormal position and malocclusion of teeth with anterior open bite.



Fig. 10-17: Fish face appearance seen in Treacher Collins syndrome.

Radiographic Features

- *Zygomatic bone*—reduction in size of zygomatic bone.
- *Maxillary sinus*—maxillary sinus is underdeveloped or completely absent (**Fig. 10-18**).
- *Articular eminence*—articular eminence is either shallow or absent.



Fig. 10-18: Underdeveloped maxillary sinus seen in cases of Treacher Collins syndrome.

- *Mastoid air cells*—mastoid air cells are reduced or absent.
- *Jaws*—hypoplasia of mandible and maxilla showing accentuation of antegonial notch and steep mandibular angle, which gives impression that the mandible is bending in an inferior and posterior direction (**Fig. 10-19**).
- *Teeth*—partial anodontia and malformation of teeth.



Fig. 10-19: Mandible is bending in inferior direction in cases of Treacher Collins syndrome.

Management

- *Cosmetic surgery*—improvement and surgical interventions should be done to improve osseous and ear defect.

Hyperplasia of Maxillary Tuberosity

There is bilateral enlargement of maxillary tuberosity. It is usually seen in adults. There is difficulty in wearing denture and normal mastication. Radiologically, bone is

more opaque than normal. Surgical removal of maxillary tuberosity can be done.

Hyperplasia of Coronoid Process

It is seen in adults, with onset at puberty. There is limited opening, which may be unilateral or bilateral. Enlargement can be seen radiologically. Surgical removal can be done.

Focal Osteoporotic Bone Marrow Defect

It is area of hematopoietic marrow which is large in size to confuse with intraosseous radiolucency.

Etiopathogenesis

- *Persistence of fetal marrow*—they are derived from bone marrow hyperplasia of persisting embryonic marrow remnants.
- *After tooth extraction*—there may be aberrant bone regeneration after tooth extraction.
- *Increased demand for erythrocyte*—marrow hyperplasia can occur in response of increased demand for erythrocyte.
- *Other factors*—it can also occur in trauma and local inflammation.

Clinical Features

- *Sex*—it is more common in females than males.
- *Common sites*—molar, premolar region in mandible.
- *Symptoms*—asymptomatic with history of pain in that region.

Radiographic Features

- *Location*—it is most common in edentulous areas.
- *Internal structure*—radiolucent area demonstrates internal trabeculation. Occasionally, trabeculae with spotty radiopaque flecks are seen within the radiolucency (Fig. 10-20).



Fig. 10-20: Focal osteoporotic bone marrow defect showing radiolucent defect with spotty radiopaque flecks.

- *Margins*—margins may be well-defined or less defined. They may be corticated or non-corticated.

Diagnosis

Diagnosis is made by exclusion.

Differential Diagnosis

- *Residual dental infection*—sign of infection is present.
- *Central neoplasm*—margins are not so corticated.

Management

There is no specific treatment for it.

Chondroectodermal Dysplasia

It is also called as 'Ellis-van Creveld disease'. The Ellis-van Creveld (EvC) syndrome was first described by Dr. Richard W.B. Ellis of Edinburgh and Simon van Creveld of Amsterdam.

It is congenital and the patient present evidence of chondrodysplasia, ectodermal dysplasia, polydactylism and congenital morbus cordis.

Clinical Features

- *Hands*—post-axial polydactyly in the hands, i.e. an extra finger lateral to the normal fifth finger, is a consistent finding. Polydactyl in the feet is a rare finding.
- *Bone dysplasia*—bone dysplasia is characterized by acromesomelia, i.e. relative shortening of the distal (acromelic) and middle (mesomelic) segments. It may interfere with the ability to make a tight fist.
- *Knees*—there may be a deformity of the knees that, frequently progress and causes a significant malalignment.
- *Nails*—the nails of the fingers and toes are dysplastic.
- *Congenital heart defect*—congenital heart defects may include hypoplasia of aorta, atrial and ventricular septal defects and a single atrium. There is dwarfism, i.e. the long bones being short and the trunk of normal length.
- *Hair*—there is absence of nails and the hair tend to be fine and sparse.

Oral Manifestations

- *Teeth*—teeth are deficient in number; and those which develop are small, rudimentary, conical, spaced and irregular in position. The teeth are affected, with eruption occurring at birth or shortly thereafter.
- *Deciduous molars*—the deciduous molars present a crenate occlusal surface. The permanent dentition is more likely to be defective than the deciduous one. Eruption is delayed.

- **Partial hairlip**—the lip deformity often referred to as a “partial hairlip” results from an abnormally short upper lip, which may also be sunken secondary to hypoplasia of maxilla.

Radiographic Features

As mentioned previously, these are shorter distal and middle phalangeal segments, in relation to the proximal phalangeal segments. Deciduous molars show typical shape.

Diagnosis

- **Clinical diagnosis**—polydactyly with hairlip and partial anodontia will favor the diagnosis of Ellis-van Creveld syndrome.

Management

- **Surgical**—management of hairlip is done by surgical method for cosmetic reason.

Arhinencephaly

It is a developmental abnormality of the skull and face in which there is absence or deficiency of the olfactory portion of brain.

Clinical Features

- **Hypotelorism**—absence of the vertical and cribriform plates of ethmoid and of crista galli result in the orbits being more closely set, a condition known as *hypotelorism*.
- **Lack of nasal bone**—there is depression of nose and absence of the bridge because of lack of nasal bones and nasal septum.
- **Prow of boat appearance**—in some cases, an associated deformity of the frontal portion of skull presents an angular appearance resembling ‘the prow of a boat’ presumably due to premature fusion of metopic sutures.

Oral Manifestation

- **Cleft lip**—there is wide cleft in the central portion of the lip, where the philtrum is absent and with failure of development of any part of the premaxilla, the cleft is continuous through the palate.

Diagnosis

- **Clinical diagnosis**—Hypotelorism with cleft lip and depressed nasal bridge will aid in diagnosis.

Management

- **Surgery**—surgery of cleft lip is done for cosmetic reasons and for functional reasons.

Phlebectasia

It is first used by Gerwig in 1928. It is an isolated, abnormal, fusiform or saccular dilatation of veins. It is also known as *venous congenital cyst*, *venous aneurysm*, *venous ectasia* or *essential venous dilatation*.

Etiology

- Mechanical compression of left innominate vein by a high tortuous aorta in hypertension or of venous structures between the sternum and the left innominate artery in pectus excavatum.

Clinical Features

- **Appearance**—it usually appears as isolated swelling. Swelling is not visible before the compression but as soon as patient clinch the jaws, swelling is visible (**Figs 10-21A and B**).



Fig. 10-21A: Without compression of neck no swelling is visible.



Fig. 10-21B: Note the swelling seen on left side of face below the ear after the compression (Courtesy Dr Abhishek Soni, Lecturer, Periodontia, VSPM Dental College and Research Center, Nagpur, India).

- *Age and sex distribution*—it is mostly seen in children/young adults on left side. Male: Female ratio is 2:1
- *Site*—commonly affects internal jugular vein, external jugular vein, anterior jugular vein and superficial communications.
- *Other features*—rarely arteriovenous fistulas or cardiac anomaly may be present.

Radiological Features

- *Ultrasonography*—dilated blood vessels are usually seen on ultrasonography (Fig. 10-22).

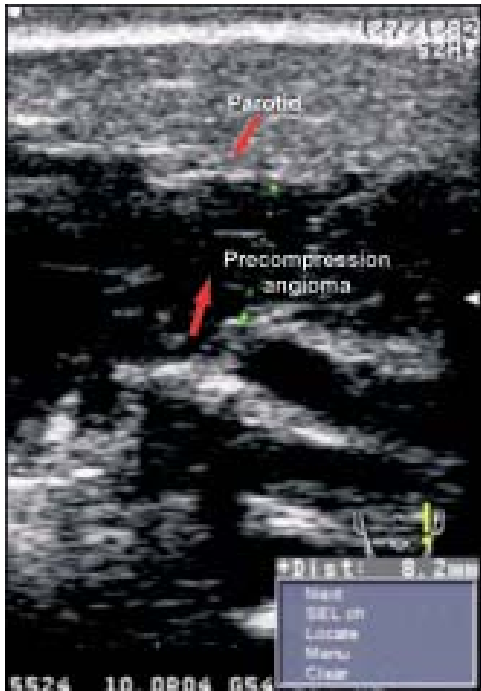


Fig. 10-22: Decompression image on sonography of phlebectasia (Courtesy Dr Abhishek Soni, Lecturer, Periodontia, VSPM Dental College and Research Center, Nagpur, India).

- *Contrast computed tomography scan*—soft tissue mass is usually seen on the CT scan. There are chances of Phleboliths formation, which can be seen in contrast CT as radiopacities (Fig. 10-23).
- *Venography*—dilated venous channels seen on venography (Fig. 10-24).

Treatment

It can be surgical, embolization or injection of sclerosing agents.

Ectodermal Dysplasia

It represents a group of inherited diseases in which two or more ectodermally derived structure does not develop. It can be autosomal dominant, autosomal recessive and

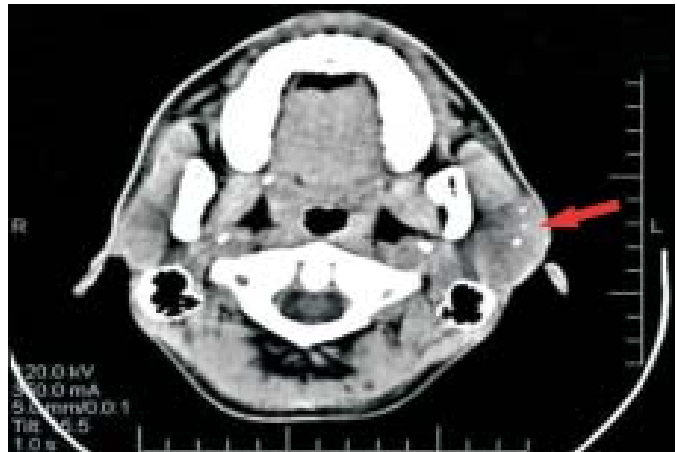


Fig. 10-23: Contrast CT picture (Courtesy Dr Abhishek Soni, Lecturer, Periodontia, VSPM Dental College and Research Center, Nagpur, India).

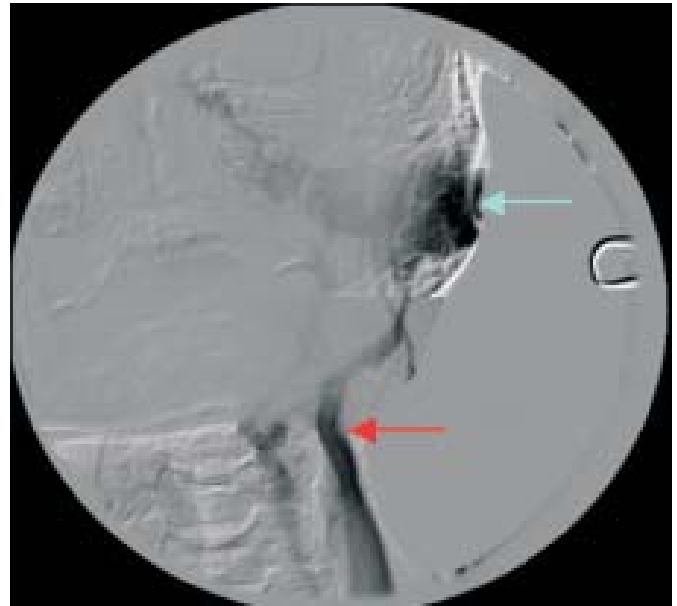


Fig. 10-24: Venography picture of jugular vein, venous dilatation (green arrow) and left jugular vein (red arrow) (Courtesy Dr Abhishek Soni, Lecturer, Periodontia, VSPM Dental College and Research Center, Nagpur, India).

X-linked. There are many types of ectodermal dysplasia but they are very rare. The most commonly occur ectodermal dysplasia is called as 'hereditary hypohidrotic (anhidrotic) ectodermal dysplasia'. This shows X-linked inheritance.

Clinical Features

- *Sex*—males are affected more frequently than females.
- *Appearance*—it is characterized by hypotrichosis, hypohydrosis and anhidrosis with saddle nose appearance.
- *Hairs*—the hair of scalp and eyebrows tend to be fine, scanty and blond.

- *Frontal bosses*—supraorbital and frontal bosses are pronounced.
- *Heat intolerance*—skin is often dry, soft, smooth and scaly with partial or complete absence of sweat glands. Such patient cannot perspire and they usually suffer from hyperpyrexia and inability to endure warm temperature (Fig. 10-25).
- *Sibling appearance*—facial appearance of these individual resemble to each other, enough to be mistaken for siblings.
- *Palate*—the palatal arch is frequently high and cleft palate may be present.
- *Salivary gland*—along with all this, since the salivary gland, including the intraoral accessory gland, are sometimes hypoplastic, they result in xerostomia and dry cracked lips with fissuring at the corners of mouth.
- *Midfacial hypoplasia*—midfacial hypoplasia results in protuberant lip of the patient. This occurs due to reduction in vertical dimension.



Fig. 10-25: Dry skin appearance of skin in patient with ectodermal dysplasia with protuberant lip.

Oral Manifestations

- *Oligodontia or anodontia*—patients with this abnormality invariably manifest oligodontia or partial absence of teeth (Fig. 10-26) with frequent malformation of any present tooth in deciduous and permanent dentition. Where some teeth are present they are commonly truncated or cone shaped.

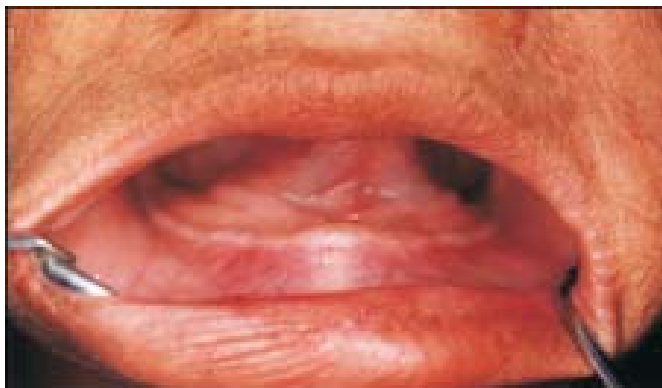


Fig. 10-26: Absence of teeth in patient with ectodermal dysplasia.



Fig. 10-27: Reduction in vertical dimensions of alveolar bone of mandible in case of ectodermal dysplasia.

Radiological Features

- *Alveolar bone*—since the alveolar process does not develop in the absence of teeth, they appear as thin and less vertical dimension (Fig. 10-27).
- *Teeth*—radiograph will also show absence of teeth (Fig. 10-28).



Fig. 10-28: Lateral view of patient showing thin alveolar bone and absence of teeth (Courtesy Dr Chole).

Diagnosis

- *Clinical diagnosis*—absence of teeth with dry skin is valuable aid in the diagnosis of ectodermal dysplasia.
- *Radiological diagnosis*—reduction in the height of alveolar bone.
- *Special impression of fingertip*—this is taken in the childhood. This impression is taken to count density of sweat gland.

Management

- *Prosthetic replacement*—from dental point of view, partial and complete dentures should be constructed for both functional and cosmetic purpose.
- *Endogenous dental implant*—it can be considered by selecting proper site in the patient older than 5 years of age.

Gardner's Syndrome

It is also called as familial multiple polyposis. It is hereditary condition. The responsible gene for this syndrome is chromosome 5.

Clinical and Radiological Features

- *Age*—it is usually noticed in second decade of life with male predilection.
- *Colonic polyps*—these are commonly found in intestine. This polyp has high degree of transformation into adenocarcinoma.
- *Osteoma*—osteoma are most common in frontal bone, mandible, maxilla and sphenoid bone (Fig. 10-29). Osteoma in mandible is present in area of mandibular angle.
- *Facial deformity*—facial deformity may occur due presence of osteoma in the mandible.

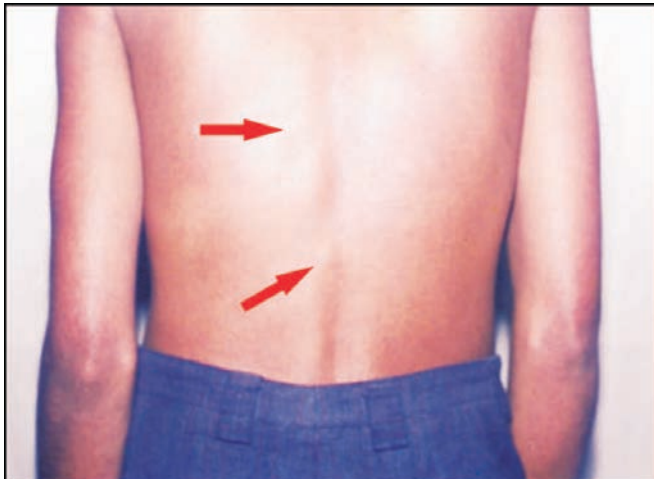


Fig. 10-29: Osteoma seen on back of patient (Courtesy Dr Datarkar, Asso. Professor, Department of Oral Surgery, SPDC, Wardha, India).

- *Limitation of opening*—in some cases of large osteoma of mandibular angle, opening of the patient mouth may be reduced.
- *Other features*—some of the patient may have epidermoid cyst, thyroid carcinoma and desmoid tumor.

Radiological Features

- *Supernumerary teeth*—there is presence of unerupted supernumerary teeth in the jaws (Fig. 10-30).
- *Osteomas of mandible*—these areas appears to be increased in radiodensity. Size varies from small to large.

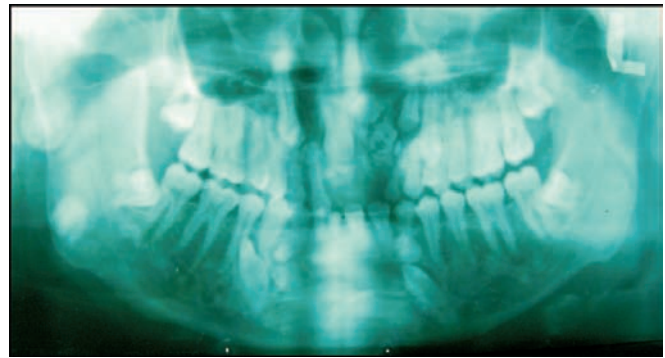


Fig. 10-30: Multiple supernumerary teeth seen in case of Gardner's syndrome (Courtesy Dr Datarkar, Asso. Professor, Department of Oral Surgery, SPDC, Wardha, India).

Diagnosis

- *Clinical and radiological diagnosis*—supernumerary teeth with osteoma in the jaw or other part of the body will favor the diagnosis of Gardner's syndrome.

Management

- *Prophylactic colectomy*—it should be done as there are high degree of transformation into malignancy.
- *Removal of jaw osteoma*—it can be removed for cosmetic reason.

Turner's Syndrome

It is also called as XO syndrome. Turner syndrome is a chromosomal condition that alters development in females. Turner's syndrome is a chromosomal condition related to the X chromosome.

Clinical Features

- *Short stature*—women with this condition tend to be shorter than average. They often have normal height for the first three years of life, but then have a slow growth rate. At puberty, they do not have the usual growth spurt (Fig. 10-31).
- *Webbed neck*—extra skin present on the neck.
- *Lymphedema*—there is also puffiness or swelling (lymphedema) of the hands and feet.

- *Other features*—there is also skeletal abnormalities, heart defects and kidney problems. Women are unable to conceive a child (infertile) because of an absence of ovarian function.
- *Middle ear infection*—in early childhood, girls who have Turner syndrome may have frequent middle ear infections. Recurrent infections can lead to hearing loss in some cases.
- *Osteoporosis*—it can develop due to lack of estrogen.



Fig. 10-31: Short stature of individual seen in Turner syndrome.

Oral Manifestation

- *Palate*—patient noticed high arch palate (**Fig. 10-32**)
- *Teeth*—eruption of teeth occurs prematurely.
- *Corner of mouth*—corners of mouth appears pulled down (**Fig. 10-33**).

Diagnosis

- *Clinical diagnosis*—webbed neck, a broad chest and short stature. Orally high arch palate with corner of mouth pull down is typical feature.

Management

- *Growth hormone injection*—growth hormone injections are beneficial in some individuals with Turner syndrome. Estrogen replacement therapy is usually started at the time of normal puberty.



Fig. 10-32: High arch palate seen in Turner's syndrome.



Fig. 10-33: Corner of mouth appear pull down in Turner's syndrome.

Kallman Syndrome

Kallman syndrome is a rare X-linked recessive disease characterized by anosmia, underdeveloped genitalia and sterile gonads.

Clinical Features

- *Sex predilection*—it affects primarily males.
- *Anosmia*—reduced or complete absence of the sense of smell. Impaired or lack of sense of smell is caused by the absence of the olfactory bulbs.
- *Underdeveloped genital*—it is apparent when they fail to begin puberty and to develop secondary sexual characteristics (**Fig. 10-34**). Kallman syndrome also affects the hypothalamus. The hypothalamus produces reduced levels of GnRH, the hormone responsible for the secretion of the hormone LH. LH is the hormone that stimulates gonadal and genital development.
- *Osteoporosis*—it can occur in some cases.



Fig. 10-34: Underdeveloped genital seen in Kallman syndrome (Courtesy Dr Rajanikant).

Oral Manifestation

- *Tooth agenesis*—it occurs more frequently.
- *Mandibular inclination*—increased mandibular inclination and mandibular angulation is also present.
- *Cleft lip*—cleft lip is also present in this syndrome. When clefting occurred, extreme retrognathism of both maxilla and mandible was seen, a deviation which worsened during growth (**Fig. 10-35**).



Fig. 10-35: Cleft lip in patient with Kallman syndrome (Courtesy Dr Rajanikant).

Management

- *Hormonal therapy*—hormone, estrogen or testosterone, replacement, and pulsatile GnRH injections should be given.

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11

Keratotic and Non-keratotic Lesions

3

Introduction

White lesion of oral mucosa comprises of a group of conditions which can appear in variety of clinical forms. A white lesion is described as abnormal area of the oral mucosa that appear whiter than the surrounding tissue and are usually slightly raised, roughened, or otherwise of a different texture from adjacent normal tissue. The white lesion may be smooth, folded, shaggy, elevated, lacy or annular. Each of these may be in turn fissured, ulcerated, eroded or inflamed and occur as solitary, multiple, focal or diffuse lesions.

Classification of lesion is described in Table 11-1.

Normal Variation

Leukoedema

It is an abnormality of the buccal mucosa, which clinically resembles early leukoplakia.

Etiology

- *Racial*—as it is prevalent in black racial cause can be present.
- *Tobacco*—it is seen more commonly in smokers than non-smokers.
- *Poor oral hygiene*—this can also predispose for the occurrence of leukoedema.

Clinical Features

- *Age, race and sex distribution*—it is common in age group of 15 to 35 year with prevalence in black. Male predilection is in the ratio of 2:1.
- *Sites*—the most common sites of involvement are buccal mucosa and lip. The lesion is bilateral in occurrence.
- *Velvet-like folded appearance*—buccal mucosa retains the normal softness and flexibility but exhibits grayish white,

Table 11-1: Classification of lesion

Normal variation

- Leukoedema
- Fordyce's granules
- Linea alba

Non-keratotic white lesions

- Habitual cheek biting
- Burns
- Uremic stomatitis
- Radiation mucositis

Candidiasis

- Thrush
- Acute atrophic candidiasis
- Chronic atrophic candidiasis
- Id reaction
- Extraoral candidiasis
- Chronic mucocutaneous candidiasis
- Systemic candidiasis

Keratotic white lesion

- Traumatic keratosis
- Keratosis associate with dental restoration
- Intraoral skin graft
- Focal epithelial hyperplasia
- Psoriasis

Oral genodermatoses

- White sponge nevus
- Hereditary benign intra-epithelial dyskeratosis
- Pachyonychia congenita
- Porokeratosis
- Keratosis follicularis
- Warty dyskeratoma
- Hyalinosis cutis et mucosa oris
- Pseudoanthoma elasticum

slightly folded opalescent appearance that is described as epithelium covered with diffuse edematous film or *velvet-like veil* (Fig. 11-1).

- *Mother of pearl appearance*—in some cases, lesion is diffuse and shows a filmy, mother of pearl appearance, often with delicate overlapping curtain-like mucosal folds.
- *Signs*—in this disease desquamation may occur which may leave surface of the lesion eroded.

- *Diagnostic test*—it can be eliminated by the stretching and scraping of mucosa but re-establishes itself almost immediately.
- *Histological*—it shows characteristic edematous cells.



Fig. 11-1: Leukoedema showing slightly folded appearance.

Diagnosis

- *Clinical diagnosis*—faint milky folded appearance which is eliminated by stretching the mucosa.

Differential Diagnosis

- *Leukoplakia*—leukoedema has faint milky appearance, folded and wrinkled pattern as compared to definite whiteness of leukoplakia. Leukoplakia cannot be eliminated by stretching.
- *Cheek biting lesion*—it is usually unilateral in appearance as compared to bilateral occurrence of leukoedema. Tissue tag is usually seen.
- *White sponge nevus*—white sponge nevus is thicker, plaque-like. It is not eliminated on stretching.
- *Hereditary benign intraepithelial dyskeratosis*—same as white sponge nevus. It has typical microscopic features.

Management

- No treatment is necessary as it has no pre-malignant potential.

Fordyce's Granules

Fordyce's granule is a developmental anomaly characterized by heterotrophic collection of sebaceous glands at various sites in oral cavity which is covered with intact mucosa. These are usually submucosal.

Pathogenesis

It has been postulated that the occurrence of sebaceous glands in the mouth may result from the *inclusion* in the oral cavity of the ectoderm having some of the potentialities of the *skin* in the course of development of the maxillary and mandibular processes during embryonic life.

Clinical Features

- *Age and sex distribution*—it is seen in any age group with somewhat more prevalent in males as compared to females.
- *Sites*—it is most commonly found bilaterally in symmetrical pattern on mucosa of the cheek, opposite to the molar teeth (Fig. 11-2). It is also found on inner surface of lips, in retromolar area lateral to anterior faucial pillar and occasionally on the tongue, gingiva, frenum and palate.
- *Symptoms*—patient usually feels roughness to hands.
- *Appearance*—they appear as small yellow spots, either discretely separated or forming relatively large plaques often projecting slightly above the surface of tissue.
- *Surface*—they are sharply delineated with smooth surface which is not ulcerated.
- *Consistency*—they have slightly cheesy consistency.
- *Plaque-like lesion*—the granules may be isolated or they may occur in confluent sheets. Sometimes they may occur in clusters and may form plaque-like lesions.
- *Tongue*—it appears as dome-shaped nodules from a few millimeters to 2 cm in diameter on the midline dorsum of the tongue.
- *Hormonal effect*—it increases rapidly in number at puberty and continues to increase throughout the adult life.
- *Histopathological*—it consists of sebaceous acini.



Fig. 11-2: Fordyce's granules seen on buccal mucosa of patient.

Diagnosis

- *Clinical diagnosis*—small yellow discrete spot separated from oral mucosa.

Management

- As such no treatment is required but if it causes disfigurement then surgical removal can be done.

Linea Alba

It is also called as '*white line*'. Linea alba refers to the line of keratinization, found on the buccal mucosa parallel to the line of occlusion expanding to a triangular area inside each labial commissures.

Etiology

- *Dietary variation*—linea alba may occur due to variation in dietary practice. The person who is taking hard diet has got more chances of having linea alba.
- *Frictional irritation*—it may occur due to contact of teeth with buccal mucosa.
- *Increases overjet*—linea alba is more prominent in people with little overjet of molars and premolars.
- *Other factors*—some other causative agents like smoking, and environmental irritants can also lead to linea alba.

Clinical Features

- *Sites*—common sites are buccal mucosa at the line of occlusion and where the mucosa overlies the bone as in hard palate and gingiva.
- *Appearance*—palate and gingiva appears whiter than the adjacent mucosa of the soft palate and alveolar gingiva.
- *Extent of line*—line extending horizontally from commissure to most posterior teeth (**Fig. 11-3**).



Fig. 11-3: Increased line of keratinization seen in buccal mucosa at the occlusal plane.

Diagnosis

- *Clinical diagnosis*—diagnosis can be made on clinical basis by observing line at the level of occlusion.

Management

- *Spontaneous regression*—in some cases spontaneous regression can occur. As this condition is asymptomatic and does not cause any esthetics or functional problems to patient no treatment is required.

Non-keratotic White Lesions

Chronic Cheek or Lip Biting

Superficial lesions produced by frequent and repeated rubbing, sucking or chewing movements that abrade the surface of a wide area of lip or cheek mucosa without producing discrete ulceration. It is also called as '*morsicatio buccarum* (*Morsicatio means morus or bite and buccarum—buccal mucosa*)'. The lesion if present on labial mucosa then it is called as '*morsicatio labiorum* and if it is on lateral border of tongue it is called as '*morsicatio linguarum*'.

Etiology

- *Neurological*—unconscious nervous habits, uncontrolled tongue thrusting and neuromuscular disorders such as tardive dyskinesia may lead to chronic biting of cheek or lip mucosa.
- *Psychological*—stress and anxiety may lead to cheek biting. Patient is aware of the habit but refuses to accept it.
- *Dental*—occlusal discrepancies, rough tooth surface may lead to chronic trauma in cheek mucosa.

Clinical Features

- *Age and sex*—it can occur at any age with women involving more commonly than the males.
- *Site*—it usually occurs on the buccal mucosa at the level of occlusion. It can also occur on lip and lateral border of tongue.
- *Appearance*—usually there is opaque white appearance which is homogenous. In some cases, there is lacerated and reddened area usually with patch of partly detached surface epithelium (**Fig. 11-4**).
- *Margins*—it may have sharply delineated borders or in some cases it may be poorly outlined. In some cases, contused margins present with transient whitish tags of necrotic tissue around the ulcer (**Fig. 11-5**).
- *Signs*—it is rough on palpation as area becomes thickened, and scarred.



Fig. 11-4: Habitual cheek biting showing patch of partly detached epithelial surface in retromolar area.



Fig. 11-5: Contused margin present with whitish tag of necrotic tissue in habitual cheek biting.

Diagnosis

- *Clinical diagnosis*—typical clinical appearance with typical location will lead to easy clinical diagnosis of the lesion.

Differential Diagnosis

- *White sponge nevus*—in cheek biting if the cause is removed the white lesion will disappear.
- *Chemical burns*—history will confirm the diagnosis of chemical burns.
- *Reaction to locally applied medicine*—history will confirm the diagnosis.

- *Candidiasis*—it is more common on tongue and white patches can be rub off.

Management

- *Psychotherapy*—small doses of diazepam 5 to 10 mg at bed time for the management of neurological disorders. But this does not give any long-term effect.
- *Acrylic guard*—this should be given to cover the facial surface of teeth and thereby restricting access to buccal and labial mucosa.

Burns

Burns cause transient non-keratotic white appearance of the mucosa which is attributed to superficial pseudo-membrane composed of coagulated tissue with an inflammatory exudate (saliva protects oral mucosa).

Types of Burns

According to severity

- *Mild burns*—mild burn causes keratotic white lesions.
- *Intermediate burns*—intermediate burns cause localized mucositis.
- *Severe burns*—severe burns coagulate the surface of the tissue and produces diffuse white lesions. If coagulation is severe, tissue cannot be scraped off easily leaving raw and bleeding painful surface.

According to cause

- *Thermal burn*—it is caused by hot food and beverages. It may occur by snow or dry ice.
- *Electrical burn*—it is caused by electrical rods.
 - *Contact burn*—it occurs when the patient is grounded and the body act as conductor with current passing through him along the path of least resistance. These burns are fatal.
 - *Arc burn*—it is sustained when arcs or sparks appear in the gap between a live wire and tissue.
- *Chemical burns*—it is caused by chemical agents.
- *Radiation burn*—this occurs in patient under radiation therapy for the management of malignancy in the oral cavity.

Thermal Burns

Clinical features

- *Sites*—anterior 1/3rd of tongue and palate.
- *Symptoms*—there is pain which lasts for short duration.
- *Appearance*—it may produce coagulation necrosis of superficial tissue that appears whitish.
- *Signs*—in some cases, there may be frank ulceration and stripping of mucosa. Red area is tender to painful, it may blanch on pressure and there is bleeding on manipulation. Surface layer of epithelium is desquamated.

- *Pizza burns*—central palatal burns, whitish gray or ulcerated lesions on the middle third of the hard palate (**Fig. 11-6**). These also present superficial necrosis and ulceration due to combination of heat of the cheese and its adhesion to blister epithelium.



Fig. 11-6: Pizza burns showing grayish lesion of hard palate.

- *Frost bite of lip or popsicle panniculitis*—it occurs due to dry ice (snow). In this, tongue and lip are most commonly affected. It occurs due to prolonged contact of ice cream and other frozen confectionaries or very cold metal, glass object, with child's lip. There is persistent swelling and redness. Epithelium becomes dry and rough than surrounding tissues.

Management

- *Pain control*—pain can be managed by systemic analgesics and topical hydrocarbon in emollient base.
- *Infection control*—there are chances that secondary infection may occur in this patient. So to prevent secondary infection antibiotics should be given.

Electrical Burns

More commonly type of burn occurs in oral cavity in which saliva acts as conducting medium. This burn can generate 3000 degree Celsius heat and causes lot of tissue destruction.

Clinical features

- *Age*—it is usually seen in children as they can accidentally chew live wire or end of an extension cord.
- *Site*—it occurs more frequently at the commissure region of the lips.
- *Symptoms*—as there is destruction of neural tissue the lesions are usually painless. There is also loss of sensory and motor function of surrounding tissues.
- *Appearance*—the clinical appearance of the burn is a gray white tissue surrounded by narrow rim of erythema.

- *Margins*—the center of the lesion may be depressed and margins are raised.
- *Signs*—tissue begins to swell within the few hours of injury, the margin of wound become irregular and the lips protrude and control of saliva is diminished. The resulting burn of the lips and sometimes of the gingiva and tongue cause destruction and necrosis of a considerable amount of tissue. Necrotic tissues slough between 2 to 4 weeks after the accident. Following sloughing, irregular ulceration can occur.
- *Tooth germ*—developing tooth germs or buds are often destroyed in the accident with permanent cosmetic disfigurement.
- *Facial asymmetry*—when surgery is not performed the opposing lips may adhere to each other with concomitant contraction and scarring and finally asymmetrical facial appearance may result.

Management

- *Surgery*—when severe tissue damage is there, surgery is recommended. If surgery is performed after healing, multiple surgical operations are required.
- *Tetanus immunization*—this should be given to prevent tetanus.
- *Antibiotics*—this should be given to prevent secondary infection. Most commonly antibiotics given are penicillin.
- *Intraoral splints*—in order to reduce the number of surgical operations and to improve oral asymmetry, removable and fixed intraoral splints have been recommended. The appliance has two ovoid acrylic posts that extend from the mouth and flare laterally. The post is extended to maintain the commissure distance equal from the midline.

Chemical Burns

Burns due to caustic chemical agents will produce coagulation necrosis of the epithelium with subsequent inflammation. It can occur in children which hold drug in oral cavity for extended period of time.

Causes

- *Aspirin and aspirin-containing compounds*—it occurs when it is kept in mucobuccal fold to relieve toothache.
- *Hydrogen peroxide*—concentration greater than 3% can cause epithelial necrosis.
- *Silver nitrate*—it is used in treatment of aphthous ulcer for pain relief. This may cause severe mucosal damage.
- *Toothache drop burns*—such drops contain creosote, guaiacol and phenol derivatives. In this phenol can cause extensive mucosal necrosis and underlying bone loss.
- *Ethyl alcohol burns*—topical application of ethyl alcohol solution which results in sloughing of the oral mucosa. Some of the mouth wash contain 25% ethyl alcohol.

- **Acid burns**—if the rubber dam placement is ineffective then acid used to etch tooth surface may come in contact with the oral mucosa.
- **Ingestion**—chemical burns of the oral cavity can result when the children by mistaken drink household chemical and with suicide attempts by ingestion of caustic material.

Clinical features

- **Symptoms**—the lesion is usually painful.
- **Appearance**—irregularly shaped, white pseudomembrane covered lesion develops (**Fig. 11-7**).
- **Signs**—gentle lateral pressure causes the white material to slide away exposing an exquisitely painful central red ulceration and more adherent patches of white material on the periphery. Diffuse border may present if the burn is more extensive.
- **Cotton roll burns (cotton roll stomatitis)**—cotton roll is used for control of moisture during dental treatment. Sometimes dry mucosa can adhere to cotton roll. While removing cotton roll from the oral cavity epithelium can also be removed along with it. This type of lesion is called as cotton roll burns or cotton roll stomatitis.



Fig. 11-7: Chemical burn showing white pseudomembrane type of lesion.

Diagnosis

- **Clinical diagnosis**—history is very important while coming to clinical diagnosis. If patient gives history of any medicament kept in oral cavity diagnosis of chemical burn can be made.

Differential diagnosis

- **Candidiasis**—in chemical burns there is acute onset, pain and focal area of involvement which is usually not a feature of candidiasis.

Management

- **Prevention**—prevention is best treatment for the chemical burn. Patient should be asked to not to keep tablet in oral cavity for longer period of time.

- **Pain control**—it should be done with a topical anesthetic like dyclonine hydrochloride. Pain relief may result from anesthetic effect of an antihistaminic mouth rinse.
- **Protective coat**—protective coat of emollient paste or hydroxypropyl cellulose film should be applied.
- **Surgical debridement**—when large area of necrosis is present surgical debridement of necroses tissue should be carried out.
- **Antibiotics**—this should be given to prevent secondary infection.

Radiation Mucositis

It is secondary to therapeutic radiation of head and neck cancer develops towards the end of the 1st week of therapy. It occurs if radiation given in excess of 3500 to 4000 rads.

Mucositis occurs when the rate of epithelial growth and repair are affected by radiation, resulting in epithelial thinning, erosion and ulceration.

Clinical Features

- **Age and sex**—most commonly found in children and elderly adults with male predilection.
- **Site**—radiation has more marked effect on rapidly proliferating epithelium and therefore, mucositis involves the non-keratinized mucosa first.
- **Appearance**—there is redness of oral mucosa followed by pseudomembrane formation with large area of oral mucosa covered with grayish, white slough alternating with areas of more severe ulceration (**Fig. 11-8**).
- **Signs**—the first sign of mucositis may be whitish appearance of the mucosa due to hyperkeratinization and intraepithelial edema or a red appearance due to hyperemia.



Fig. 11-8: Radiation mucositis occur due to radiation. Note severe ulceration with pseudomembrane formation on oral mucosa.

- *Acute reaction*—acute reaction occurs during the course of radiotherapy due to direct tissue toxicity. Ulcer resolves over several weeks following the completion of therapy.
- *Chronic reaction*—chronic complication or late radiation reaction occurs due to change in the vascular supply, fibrosis in connective tissue and muscle and change in cellularity of tissues.

Diagnosis

- *Clinical diagnosis*—severe mucosal ulceration with history of radiation will diagnose the condition of radiation mucositis.

Management

- *Mouth rinse*—a soothing mouth rinse such as an antihistaminic with kaopectate will offer pain relief.
- *Good oral hygiene*—it is the most effective treatment for complication of radiation therapy. Once radiation therapy is initiated, all foci of infection from oral cavity should be removed.
- *Betamethasone mouth wash*—high dose of betamethasone mouth wash is very effective treatment for radiation mucositis.
- *Other therapy*—other therapy like topical allopurinol, antimicrobial lozenges, benzydamine, capsaicin, chamomile, dyclonine HCL, milk of magnesia are tried but there is limited success for this therapy.

Uremic Stomatitis

Non-keratotic white lesions caused due to elevated creatinine or blood urea nitrogen. It has been suggested to be consequence of strongly alkaline saliva due to ammonia formation from retained urea secreted in the saliva. Most of the cases of uremic stomatitis occur in acute renal failure.

Types

- *Type I*—generalized or localized erythema with pseudomembrane formation. After removal of pseudomembrane no ulceration is present.
- *Type II*—this is severe form and after removal of pseudomembrane ulceration is present.

Clinical Features

- *Age and sex*—it is common in young and middle aged adults with no sex predilection.
- *Site*—it is predominately seen in buccal mucosa, tongue and floor of mouth.
- *Symptoms*—there is ammoniacal odor to breath. Patient may complain of unpleasant taste and burning sensation in oral cavity.

- *Appearance*—there is extensive pseudomembranous white lesion (**Fig. 11-9**).
- *Signs*—in some cases, there is appearance of ulcers.
- *Shape and size*—oral ulceration varies in size and is irregular in shape and is usually shallow.
- *Oral hairy leukoplakia*—in some cases uremic stomatitis can mimic oral hairy leukoplakia.



Fig. 11-9: White pseudomembrane lesion in patient with acute renal failure seen on buccal mucosa.

Diagnosis

- *Clinical diagnosis*—patient with uremic ulcer will manifest the clinical signs of the uremic syndrome, an ammonia odor of the breath, weight loss, anemia and cardiac arrhythmias confirms the diagnosis of uremic stomatitis.

Management

- *Palliative oral rinse*—ulcer, if painful may be treated by prescribing a palliative oral rinse such as an antihistamine oral suspension with kaopectate.
- *Acidic mouth rinse*—mild acidic mouth rinse like diluted hydrogen peroxide can clear oral lesion.

Candidiasis

It is also called as 'candidosis'. An older name for this disease is 'moniliasis'. Candidiasis is the disease caused by infection with yeast-like fungus *Candida albicans*. Oral involvement is probably the most common manifestation of human candidal infection. It can occur either solely confined to the oral mucosa or as a part of any of the several mucocutaneous candidiasis syndromes.

Classification

There are many classifications for Candidiasis (Tables 11-2 to 11-4). Some of them are listed below. Classifications are made on the basis of clinical presentation, location, etc.

Causative Organisms

It is caused by *Candida albicans* (the yeast-like fungus occurs in yeast and mycelial forms). Other candida species which can cause candidiasis are *Candida stellatoidea*, *Candida tropicalis*, *Candida parapsilosis*, *Candida pseudotropicalis*, *Candida famata*, *Candida rugosa*, *Candida krusei* and *Candida guilliermondii*.

It is present in two forms, i.e. dimorphisms. Yeast form and hyphal form.

Candida albicans is the commonest pathogen of all candida species and it appears as moist creamy colonies and on blood agar as dull gray colonies.

Predisposing Factors (Table 11-5)

- *Changes in oral microbial flora*—marked changes in oral microbial flora may occur owing to administration of antibiotics (broad spectrum), excessive use of antibacterial mouth rinses, xerostomia secondary to anticholinergic agent or salivary gland disease. This

Table 11-2: First classification

Oral candidiasis

Acute

- Acute pseudomembranous candidiasis (thrush)
- Acute atrophic candidiasis (antibiotics sore mouth)

Chronic

- Chronic atrophic candidiasis
 - Denture stomatitis
 - Median rhomboid glossitis
 - Angular cheilitis
- Id reaction
- Chronic hyperplastic candidiasis

Chronic mucocutaneous candidiasis

- Familial CMC
- Localized CMC
- Diffuse CMC
- Candidiasis endocrinopathy syndrome

Extraoral candidiasis

- Oral Candidiasis associated with extraoral lesions orofacial and intertriginous sites (candidal vulvovaginitis, intertriginous candidiasis)
- Gastrointestinal candidiasis
- Candida hypersensitivity syndrome

Systemic candidiasis

- Mainly affect the eye, kidney and skin

Table 11-3: Second classification

Primary oral candidiasis

- *Acute form*
 - Pseudomembranous
 - Erythematous
- *Chronic form*
 - Hyperplastic
 - Erythematous
 - Pseudomembranous
- *Candida associated lesion*
 - Denture stomatitis
 - Angular stomatitis
 - Median rhomboid glossitis
- *Keratinized primary lesion super-infected with candida*
 - Leukoplakia
 - Lichen planus
 - Lupus erythematosus

Secondary candidiasis

- Oral manifestations of systemic mucocutaneous candidiasis (as a result of diseases such as thymic aplasia and candida endocrinopathy syndrome).

Table 11-4: Third classification

Candidiasis of nails and skin

- Candidal onychia
- Candidal paronychia

Candidiasis confined to skin

- Interdigital candidiasis
- Intertriginous candidiasis
- Candidids (monillids)

Candidiasis confined to mucosae

Oral mucosa

- *Acute oral candidiasis*
 - Acute pseudomembranous candidiasis (thrush)
 - Acute atrophic candidiasis (antibiotics sore mouth)
- *Chronic oral candidiasis*
 - Chronic atrophic candidiasis (denture sore mouth)
 - Chronic hyperplastic candidiasis

Gastrointestinal mucosa

- Pharyngeal candidiasis
- Esophageal candidiasis
- Intestinal candidiasis

Respiratory mucosa

- Bronchial candidiasis

Genitourinary mucosae

- Candidal vulvovaginitis

Mucocutaneous candidiasis

Confined to mucocutaneous surface

- *In condition with major immunologic defect*
 - Swiss-type agammaglobulinemia
 - Hereditary thymic dysplasia
 - Di George syndrome
 - AIDS
- *In condition with minor immunological or other systemic defect*
- *Chronic mucocutaneous candidiasis (CMC) syndromes*
 - Familial mucocutaneous candidiasis
 - Candidiasis endocrinopathy syndrome
 - Localized chronic mucocutaneous candidiasis
 - Diffuse chronic mucocutaneous candidiasis
 - Chronic mucocutaneous candidiasis in association with thymoma

Confined to mucocutaneous junctions

- Candidal angular cheilitis
- Perianal candidiasis

Systemic candidiasis

- Candidal endocarditis
- Candidal septicemia
- Candidal meningitis

Table 11-5: Conditions associated with increased vulnerability of oral candidiasis and their mechanism

Category	Condition	Mechanism
Altered local resistance to infection	Poor oral hygiene Xerostomia Recent antibiotics treatment Dental appliance	Promotes organism adherence and colonization Absence of antimicrobial and flushing effect of saliva Inhibits competitive oral bacteria Isolated mucosa from saliva and functional cleansing serve as organism reservoir
Compromised immune system function	Early infancy Genetic immune deficiency AIDS Corticosteroids therapy Pancytopenia	Immune competence has not completely developed Specific humoral or cellular immune defects Deficient cellular immune response Inhibition of immune function Depletion of circulating leukocytes caused by chemotherapy, aplastic anemia and similar hemopoietic disorders
Generalized patient debilitation	Anemia, malnutrition, malabsorption Diabetes mellitus Advanced systemic disease	Epithelial thinning and altered maturation, poor tissue oxygenation Recurring hyperglycemia and mild ketoacidosis Metabolic toxicity or limited blood perfusion of tissue

change will result in inhibition of competitive bacteria resulting in candidiasis.

- *Local irritant*—chronic local irritant (denture, orthodontic appliance and heavy smoke).
- *Drug therapy*—administration of corticosteroids, cytotoxic drugs, immunosuppressive agents and radiation to head and neck.
- *Acute and chronic disease*—acute and chronic disease such as leukemia, lymphoma, diabetes and tuberculosis.
- *Malnutrition states*—malnutrition states such as low serum vitamin A, pyridoxine and iron levels.
- *Age*—age (infancy, pregnancy, old age), hospitalization and oral epithelial dysplasia.
- *Endocrinopathy*—endocrinopathies such as hypoparathyroidism, hypothyroidism and Addison's disease.
- *Immunodeficiency states*—Primary and acquired immunodeficiency state such as hypogammaglobulinemia.
- *Others*—tight and close fitting garments encourage the growth of candida. Areas around the indwelling catheter are also involved.

Thrush or Pseudomembranous Candidiasis

It is the prototype of oral infection caused by yeast-like fungus. It is the superficial infection of upper layer of oral mucous membrane and results in formation of patchy white plaque or flecks on mucosal surface.

Pathogenesis

- *Overgrowth of yeast*—overgrowth of yeast on the oral mucosa leads to desquamation of epithelial cells and accumulation of bacteria, keratin, and necrotic tissue.
- *Formation of pseudomembrane*—this debris combines to form a pseudomembrane, which may adhere closely to the mucosa.

Clinical Features

In infants

- *Age*—in neonates, oral lesions start between the 6th and 10th day after birth.
- *Cause*—infection is contracted from the maternal vaginal canal where *Candida albicans* flourishes during the pregnancy.
- *Appearance*—the lesions in infants are described as soft white or bluish white, adherent patches on oral mucosa which may extend to circumoral tissue.
- *Symptoms*—they are painless and noticed on careful examinations. They may be removed with little difficulty.

In adult

- *Sites*—common sites are roof of the mouth, retromolar area, and mucobuccal fold. But it is common on any other mucosal surface.
- *Sex*—it is common in women as compared to male.
- *Prodromal symptoms*—prodromal symptom like rapid onset of bad taste may be there. Spicy food will cause discomfort.
- *Symptoms*—patient may complain of burning sensation.
- *White plaques*—pearly white or bluish white plaques are present on oral mucosa. They resemble cottage cheese or curdled milk (**Figs 11-10 and 11-11**). Patches are loosely adherent to oral mucosa.
- *Composition of plaques*—it is composed of tangled mass of hyphae, yeast, desquamated epithelial cells and debris.
- *Adjacent mucosa*—Mucosa adjacent to it appears red and moderately swollen.
- *Wiping of patches*—white patches are easily wiped out with wet gauze which leaves either a normal or erythematous area or atrophic area (**Fig. 11-12**). This area may be painful. Deeper invasion by the organism leaves an ulcerative lesion upon the removal of patch.



Fig. 11-10: White plaque resembling as curdled milk present on lip (Courtesy Dr Chole).



Fig. 11-11: Thrush present as white plaque on the tongue.



Fig. 11-12: Red atrophic mucosa seen in patient after adherent patches are removed (Courtesy Dr Chole).

- *Inconspicuous lesion*—in some cases lesions are relatively inconspicuous (not noticeable easily) (Fig. 11-13).
- *Malignant association*—it is occasionally associated with (coexist with) dysplastic or carcinomatous change.



Fig. 11-13: Candidiasis present on the tongue as a diffuse white lesion which is not noticed easily.

Diagnosis

- *Clinical diagnosis*—pseudomembranous lesion which can be scraped off will diagnose candidiasis.

Differential Diagnosis

- *Plaque form of lichen planus*—lesions of thrush can be wiped with the help of gauze.
- *Leukoplakia*—history of recent administration of antibiotics will favor the diagnosis of candidiasis.
- *Genodermatoses*—cytological smear should be taken to confirm the diagnosis.
- *Gangrenous stomatitis*—pseudomembrane is dirty in color and not raised above the surface.
- *Chemical burns*—the superficial white material burns of oral mucosa appears thin and delicate as compared to pseudomembranous candidiasis.

Acute Atrophic Candidiasis

It is also called as '*antibiotics sore mouth*'. When the white plaque of pseudomembranous candidiasis is removed, often red atrophic and painful mucosa remains. It may arise in acute pseudomembranous candidosis after the white plaques have shed yet the infection persists, AIDS patients, in patient receiving prolonged drug therapy: topical steroids or broad spectrum antibiotics and most commonly related to wearing dentures.

Clinical Features

- *Sites*—it can occur at any site but it usually involves the tongue and tissue underlying the prosthesis.

- **Appearance**—in this type, lesion appears as red or erythematous (**Fig. 11-14**) rather than white, thus resembling the pseudomembranous type in which white membrane has been wiped off.
- **Symptoms**—patient usually described vague pain or a burning sensation. If the discomfort is not spontaneous, pain can be elicited by mild abrasive pressure with cotton gauze.
- **Signs**—careful examination reveals a few white thickened foci that rub off leaving a painful surface. Lesion closely resembles erosive lichen planus and erythroplakia.



Fig. 11-14: Acute atrophic candidiasis involving the tongue showing erythematous area (Courtesy Dr Chole).

Diagnosis

- **Clinical diagnosis**—erythematous area with condition causing diminished host resistance will diagnose acute atrophic candidiasis.

Differential Diagnosis

- **Chemical burn**—focal white area that rub off and underlying condition of diminished host resistance favors candidiasis. History of medicament is present.
- **Drug reaction**—identification of condition causing diminished host resistance is reliable differential diagnostic feature of atrophic candidiasis.
- **Syphilitic mucus patches**—discrete small white necrotic lesions on tongue, palate or lip while candidiasis is diffuse. Skin lesion of syphilis is also present.
- **Necrotic ulcer and gangrenous stomatitis**—ulcer is deeper than candidiasis.
- **Traumatic ulcer**—history of trauma is present.

Chronic Hyperplastic Candidiasis

It is also called as '*candidal leukoplakia*' because of its firm

presentation as firm and adherent white patches occurring in the oral mucosa.

Clinical Features

- **Age and sex**—it predominantly occurs in men of middle age or above. The majority of these patients are heavy smokers.
- **Sites**—it occurs on cheek, lip and tongue.
- **Appearance**—candidal leucoplakia is extremely chronic form of oral candidiasis in which firm, and white leathery plaques are found (**Fig. 11-15**).
- **Symptoms**—lesions may persist without any symptoms for years.



Fig. 11-15: Chronic hyperplastic candidiasis showing firm white appearance.

- **Signs**—It does not rub off with lateral pressure. Lesion range from slightly white to dense white (**Fig. 11-16**) with cracks and fissures occasionally present. The borders are often vague, which mimic the appearance of epithelial dysplasia. It may occur as a part of chronic mucocutaneous candidiasis.

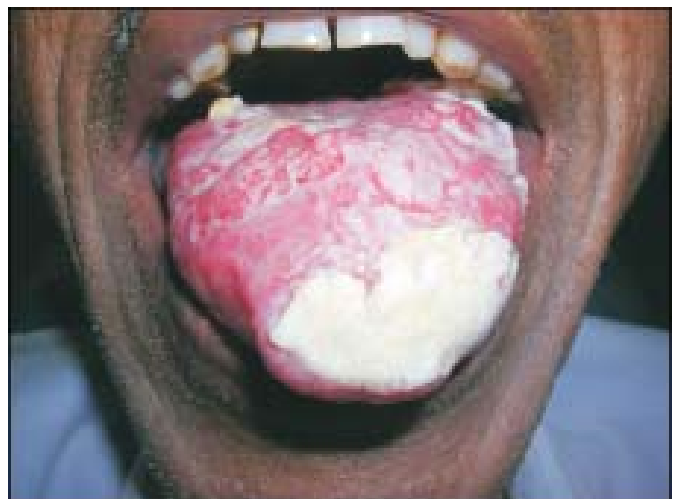


Fig. 11-16: Hyperplastic candidiasis showing fissure and crack on the tongue (Courtesy Dr Bhaskar).

Diagnosis

- *Clinical diagnosis*—a firm white leathery appearance seen which cannot rubbed off.

Differential Diagnosis

- *Lichen planus*—characteristic striated appearance, the presence of skin lesion confirm the diagnosis of lichen planus.
- *Hairy leukoplakia*—hyperplastic candidiasis improve with antimycotic treatment while hairy leukoplakia remains unchanged or progresses.
- *Superficial bacterial infection*—microscopic culture.

Id Reaction

A person with chronic candida infection may develop secondary response characterized by localized or generalized sterile vesicopapular rash that is believed to be allergic response to candida antigen (also called as monolids).

Candida Associated Lesion

Denture Stomatitis

It is also called as '*chronic atrophic candidiasis*'. It is common clinical manifestation of erythematous candidiasis. *Candida albicans* is always found in this lesion but the typical white patch of thrush does not usually develop in it. It occurs due to tissue invasion but organism effect of fungal toxin hypersensitivity to fungus.

Clinical features

- *Site*—it is usually found under complete denture or partial denture and found mostly in women and always include palate.
- *Appearance*—it exhibits patchy distribution often associated with speckled curd-like white lesion.
- *Symptoms*—soreness and dryness of mouth.
- *Signs*—palatal tissue is bright red and somewhat edematous and granular. Red patches may be erythematous or speckled. The redness of mucosa is rather sharply outlined and restricted to the tissue actually in contact with the denture (**Fig. 11-17**). The multiple pinpoint foci of hyperemia usually involving the maxilla frequently occurs.

Diagnosis

- *Clinical diagnosis*—an erythematous area under the complete denture will aid in diagnosis of candidiasis.

Differential diagnosis

- *Allergic reaction due to denture base*—it is very rare condition. If there is failure to respond to antifungal therapy then one should suspect allergic reaction.



Fig. 11-17: Denture stomatitis presented as erythematous area in maxillary area.

- *Erosive lichen planus*—skin lesion are present.
- *Dermatitis herpetiformis*—a cytological smear with periodic acid Schiff stain will disclose mycelia.

Management

- Troches containing clotrimazole and Nystatin 4 to 5 times should be applied on the denture after meal and bed time.

Median Rhomboidal Glossitis

There is debate that it is form of chronic atrophic candidiasis but area of anterior 2/3 of tongue affect by median rhomboidal glossitis is frequently followed by candida infection. It is described in detail in Chapter 22: Disease of Tongue.

Angular Cheilitis

It is described in Chapter 23: Diseases of Lip.

Treatment of Oral Candidiasis

Oral candidiasis may be treated either topically or systemically. Treatment should be maintained for 7 days. Response to treatment is often good; oral lesions and symptoms may disappear in a fairly short period (ranging from 2 to 5 days), but relapses are common because of the underlying immunodeficiency. As with other causes of oral candidiasis, recurrences are common if the underlying problem persists:

- *Removal of the causes*—replacement of denture or relining or adding mycostatin suspension below it while insertion in mouth in case of angular cheilitis and denture sore mouth. The denture must be cleaned thoroughly and regularly and should be left out of the mouth at night in hypochlorite solution. Withdrawal or change of antibiotics can be done if feasible.

- **Topical treatment**—topical treatments are preferred because they limit systemic absorption, but the effectiveness depends entirely on patient compliance. Following are most commonly used topical treatment.
 - **Clotrimazole**—it is an effective topical treatment (one oral troche [10-mg tablet]) when dissolved in the mouth five times daily. Used less frequently, one vaginal troche can be dissolved in the mouth daily. This drug has got antibacterial as well as antifungal properties.
 - **1% gentian violet** can be used but it is not ideal because of the superficial necrosis of mucosa and it may produce unsightly staining.
 - **Nystatin preparations**—it includes a suspension, a vaginal tablet, and an oral pastille. Nystatin vaginal tablets (one tablet, 100,000 units, dissolved in the mouth three times a day). Nystatin oral pastille (available as a 200,000-unit oral pastille, one or two pastilles dissolved slowly in the mouth five times a day). Nystatin oral suspension 100,000 units/cc, 1 teaspoon of which is mixed with $\frac{1}{4}$ cup of water and used as oral rinse.
 - **Amphotericin B**—another therapeutic choice is Amphotericin B (0.1 mg/ml). 5 to 10 ml of oral solution is used as a rinse and then expectorated three to four times daily. Elixir containing both tetracycline and amphotericin B may also prove to be beneficial in acute atrophic candidiasis. Addition of triamcinolone acetonide with nystatin can be used in case of angular cheilitis.
 - **Mycostatin cream**—1 lack unit or lactose containing vaginal tablet keeps under the tongue. The addition of absorbable corticosteroids and antibiotic agents to mycostatin cream and ointment accelerate the symptomatic effect. Mycostatin can be used as rinse for 7 to 10 days 3 to 4 times a day.
 - **Idoquinol**—it has antifungal and antibacterial properties. When this is combined with corticosteroid is very helpful in management of angular cheilitis.
- **Systemic treatment**—several agents are effective for systemic treatment.
 - **Nystatin**—it is polyene agents given 250 mg TDS for 2 weeks followed by 1 troche per day for third week. Nystatin has bitter taste, so to avoid patient discomfort. Sucrose or flavoring agents should be added.
 - **Ketoconazole**—it is imidazole derivative and taken as 200-mg tablet with food once daily. Patient compliance is usually good. Careful monitoring of liver function is necessary for long-term use because of reported side effects, including hepatotoxicity. Lack of efficacy of ketoconazole may occur as it requires acidic environment for proper absorption.

- **Fluconazole**—it is a triazole antifungal agent effective in treating candidiasis (100-mg tablet taken once daily for 2 weeks). Several studies suggest fluconazole is effective as a prophylactic agent, although the most effective prophylaxis dosing regimen is still unclear. Numerous reports, however, describe oral and esophageal candidiasis failing to respond to treatment with fluconazole, and in some of these cases investigators isolated resistant strains.
- **Itraconazole**—it (100-mg capsules) may be used for the treatment of oral candidiasis (200 mg daily orally for 14 days). Itraconazole oral suspension is now available (200 mg daily for 2 weeks). Salivary levels of itraconazole are maintained for several hours after administration. Itraconazole is contraindicated in patient taking astemizole, triazolam, midazolam and cisapride.

Chronic Mucocutaneous Candidiasis

Oral candidiasis may also be seen in immunological group of disorders called as mucocutaneous candidiasis. Autosomal recessive pattern is observed in this type of candidiasis. Oral lesions in this are usually thick white plaques which are not rub off.

It is caused by defect in cellular immunity and defect in structure of epidermis. Other factors which are responsible for chronic mucocutaneous candidiasis are diabetes, steroid therapy and in some cases of pregnancy.

Types

- **Chronic familial mucocutaneous candidiasis**—it is an inherited disorder, probably an autosomal recessive disorder and affects both sexes.
- **Chronic localized mucocutaneous candidiasis**—this form occurs early in life with oral mucosal, skin and nail involvement.
- **Endocrinopathy candidiasis syndrome**—in this there is subsequent appearance of hypoparathyroidism, hypoadrenocorticism and other endocrine abnormalities.
- **Chronic diffuse mucocutaneous candidiasis**—lesions involve pharyngeal mucosa and it is diffuse in nature.
- **Chronic mucocutaneous candidiasis in association with thymoma**—rarely patient with thymoma manifest chronic mucocutaneous candidiasis.

Clinical Features

- **Chronic familial mucocutaneous candidiasis**—it is characterized by candidiasis of mouth, nails, and skin. Children under the age of 10 years show superficial candidal infection. Oral candidiasis is chronic hyperplastic exhibiting firm white patches involving either buccal or lingual mucosa.

- *Chronic localized mucocutaneous candidiasis*—there is presence of chronic oral candidiasis and hypoplastic infection of nails fold in infancy. There are white patches characterized by horny masses mainly found on face and scalp. Patient show increase tendency to fungal and bacterial infection in the absence of immunological or genetic defect.
- *Candidiasis endocrinopathy syndrome*—it is also genetically transmitted, characterized by early onset and candida lesion of skin, scalp, nails and mucous membrane. There is occurrence of dental hypoplasia and severe caries. There may be balanitis and vulvovaginitis, keratoconjunctivitis, alopecia and juvenile cirrhosis.
- *Chronic diffuse mucocutaneous candidiasis*—it is of late onset and exhibits extensive raised crusty sheets involving the limbs, groin, face, scalp and shoulders. Mucocutaneous candidiasis is characterized by extensive oral, pharyngeal and often laryngeal involvement. Cutaneous involvement is characterized by raised, crusty proliferative lesions predominately on the skin of face and scalp. These lesions are also called as 'candidal granuloma'. Oral lesions are widespread, proliferative and raised white lesions.
- *Chronic mucocutaneous candidiasis in association with thymoma*—Oral and skin surface are usually involved.

Management

- *Antifungal drugs*—for resistance form of CMC and for systemic candidiasis miconazole (250 mg 6 hourly), 5-flucytosine (200 mg daily) in addition to Amphotericin B is available.
- *Transfusion of transfer factor*—transfusion of transfer factor and transplantation of culture of thymic fragments have been used to correct T. cell deficiency associated with CMC.

Systemic Candidiasis

This term is used when there is presence of both necrotizing inflammation and candida granuloma formation in one or more visceral organ usually as a result of hematogenous dissipation of organism.

Etiology

- *Immunosuppressive patient*—it occurs in immunosuppressive patient as a result of leukemia, lymphoma, steroids, cytotoxic therapy and extensive therapy.
- *Contaminated instrument*—contaminated IV instruments and indwelling catheter may lead to systemic candidiasis.
- *Drugs*—use of contaminated drug, paraphernalia by drug abuses.

- *AIDS*—this is immunological disorder in which systemic candidiasis may occur.

Clinical Features

- *Candidal endocarditis*—it is characterized by fever, dyspnea and edema of congestive cardiac failure. Vascular vegetations of candidal growth often result in embolization to major vessels. The disease is fatal in majority of cases.
- *Candidal meningitis*—it is predominately disease of male children characterized by variable features ranging from stiffness of the neck, hemiplegia and other neurological signs. The finding of Candida in spinal fluid is of diagnostic importance. The condition is fatal in half of the cases.
- *Candidal septicemia*—it occurs in those with severe oral and esophageal thrush. Features include fever, chills, shock and coma. Condition can be fatal if not treated in time.

Management

- Systemic candidiasis requires systemic administration of high doses of *Amphotericin-B* 10 mg QID, *miconazole* and *5-flucytosine*. *Mycostatin* 50,000 unit 8 hourly. Daily use of 200 mg *ketoconazole* for 2 weeks. Side effects are increased liver enzyme, abdominal pain.

Gastrointestinal Candidiasis

An extension of oral infection may occasionally lead to either pharyngeal or esophageal involvement.

Causes

- *Drugs*—long term broad-spectrum antibiotics, corticosteroid therapy and immunosuppressive drugs.
- *Diseases*—leukemia, diabetes and acquired immunodeficiency syndrome.

Clinical Features

- *Symptoms*—it presents acute enterocolitis, diarrhea, as proctitis with anal pruritis of perineal eczematation. Dysphagia, chest pain, and GIT bleeding may be presenting symptoms.
- *Esophageal candidiasis*—esophageal candidiasis with or without gastric ulceration are common forms of GIT candidiasis.
- *Appearance*—it is diagnosed by characteristic appearance of edematous ulcerated mucosa on barium swallow.

Management

- The use of topical and systemic antifungal agents.

Candidal Onychia and Paronychia

It is the candidal infection of nail and of the soft tissue on the sides and at the base of the nail and is common in housewives, nurses, and dishwasher, bartenders, and fruit pickers.

Involved nail show green black discoloration and transverse ridges. Soft tissues surrounding the nails are inflamed and tender.

Treatment is by daily topical application of amphotericin B lotion or nystatin ointment.

Interdigital Candidiasis

It is also called as *erosion interdigitalis*. It commonly involves 3rd and 4th fingers.

It is characterized by pruritic, inflamed, and scaling lesions between fingers. Treatment includes topical application of amphotericin B lotion or nystatin ointment.

Intertriginous Candidiasis

It is the candida infection of skin surface such as waist, groin, armpit, elbows, submammary area, gluteal fold, axilla, and scrotum. Intertriginous candidiasis causes colonization in skin like axilla or submaxillary fold.

Opposing skin surface in these sites, particularly in obese person prevent adequate ventilation and remain moist favoring candidal growth.

Candida has characteristic way of causing dry pustulation with desiccation in lower level of stratum corneum of skin. Lesions are very tender. Lesion spread from affected skin as an area of glaze red skin or an easily detached overlying epidermis i.e. often invaded leaving paper-like fungal lesion along the margin. Satellite lesion may develop around deeper denuded tissue.

Bronchial Candidiasis

It is a chronic infection of bronchial mucosa which may last for years without producing severe discomfort. It mimics chronic bacterial infection of the bronchus and chronic cough with mucoid sputum is common symptoms experienced by the patients.

Diagnosis is established by the demonstration of candida in sputum and through culture studies.

Management includes systemic use of antifungal agents.

Candidal Vulvovaginitis and Balanitis

Candida vulvovaginitis is a common form of candidiasis, which increases during pregnancy, diabetes, and uses of antibiotics. Candidal inflammation of glans penis may occur due to sexual contact. Diabetic women and those on long term drug therapy are prone to develop vaginal infection.

It is characterized by erythematous plaques on the glans penis and around the prepuce. Symptoms usually clear up on topical application of antifungal agents.

Candidiasis with Major Immunological Defect

- *Swiss-type agammaglobulinemia*—it is characterized by severely impaired resistance to infections of all types as a result of failure of thymic development and antibody production. Infants die in the first year of life. Oral candidiasis is a consistent feature of this syndrome.
- *Hereditary thymic dysplasia*—impairment of cell mediated immunity due to thymic dysplasia favors severe viral and candidal infection. Oral candidiasis is predominant feature of this disease.
- *Di George syndrome*—it is characterized by under-developed thymus and parathyroid glands due to a defective development of the 3rd and 4th branchial pouches. Oral candidiasis is a consistent feature of this disease.
- *AIDS*—oral manifestation of AIDS among other features include candidiasis of the oral mucosa, where the candidal infection is opportunistic in nature.

Keratotic White Lesions with no Definite Precancerous Potential

Traumatic Keratosis

It refers to an isolated area of thickened whitish oral mucosa that is clearly related to identifiable local irritant and resolves following elimination of irritant.

Etiology

- *Local irritants*—ill fitting denture, sharp clasp and rough edges of restoration are responsible for traumatic keratosis.
- *Cigarettes smoking*—heavy cigarettes smoking may also lead to this type of lesion.

Clinical Features

- *Sites*—most common sites are lip and buccal mucosa.
- *Appearance*—there is isolated thickened whitish area (**Fig. 11-18**).
- *Glassblower's white patch*—it is variant of traumatic keratosis affecting the cheek and lips, which occur in glass factory.

Diagnosis

- *Clinical diagnosis*—isolated white patch with identified local irritant will diagnose the traumatic keratosis.



Fig. 11-18: Isolated white patch seen on buccal mucosa due to sharp cusp.

- *Laboratory diagnosis*—there is varying degree of hyperkeratosis, parakeratosis and acanthosis.

Management

- *Removal of local irritant*—upon removal of the offending agent, the lesion should resolve within 2 weeks.
- *Biopsy in cases of unhealed lesion*—biopsies should be performed on lesions that do not heal to rule out a dysplastic lesion.

Psoriasis

It is a common dermatological disease characterized by white, scaly papules and plaque on an erythematous base that preferentially affects the extremities and scalp. It is characterized by increased activity of keratinocytes.

Etiology and Precipitating Factors

- *Hereditary*—it has been suggested a possible inheritance pattern, possibly transmitted as simple dominant trait.
- *Infection*— β -hemolytic streptococcal infection often precedes psoriasis.
- *Drugs*—antimalarials, β -blocker and lithium may worsen psoriasis and the rash may 'rebound' after stopping systemic corticosteroids or potent local corticosteroid.
- *Emotion*—anxiety precipitates some exacerbation.
- *Neurogenic factor*—mental anxiety and stress can increase severity of the disease.

- *Trauma*—in areas of skin damage such as scratches or surgical wound.
- *Others*—metabolic disturbances and endocrine dysfunction can also precede psoriasis.

Types (Clinical)

- *Stable plaque psoriasis*—it is most common type. The lesions are red with dry, silvery-white scaling, which may be obvious only after scraping the surface.
- *Guttate psoriasis*—it is seen in children adolescents and may follow streptococcal sore throat. Individual lesions are droplet-shaped, small and scaly.
- *Erythrodermic psoriasis*—the skin becomes universally red and scaly or more rarely just red with very little scale present.
- *Pustular psoriasis*—it is a severe form of psoriasis with eruption of minute pustule with shedding of nails is common.

Clinical Features

- *Age*—it commonly affects adults and arises in 2nd and 3rd decades of life.
- *Sites*—it commonly occurs on extremities, and scalp.
- *Progress*—it is usually chronic with acute generalized exacerbations. It is more severe in winter and less severe in the summer as a result of increase exposure to ultraviolet light.
- *Symptoms*—in some cases, patient may complain of itching.
- *Appearance*—it is characterized by occurrence of small sharply defined, dry papules each covered by delicate silvery scale which appear as a thin layer of mica (Fig. 11-19).
- *Margins and shape*—papules are enlarged at periphery and may form large plaques which are roughly symmetrical (Fig. 11-20).
- *Signs*—after removal of scale the surface of skin is red and dusky in appearance.
- *Auspitz's sign*—if the deep scales are removed one or more bleeding points are seen.
- *Psoriatic arthritis*—this is the complication of psoriasis which can involve the temporomandibular joint.

Oral Manifestations

- *Sites*—oral lesions are reported on lip, buccal mucosa, palate, gingiva and floor of mouth.
- *Appearance*—they appear as plaques, silvery, scaly lesions with an erythematous base.
- *Signs*—sometimes they are multiple papular eruptions which may be ulcerated or as small, papillary elevated lesions with scaly surface.



Fig. 11-19: Thin layer of mica appearance seen in psoriasis patient.



Fig. 11-20: Sharply defined papules seen on face of the patient (Courtesy Dr Parate).

- Usually four types of oral manifestations occur in patient with psoriasis:
 - *The first type* consists of minute well defined gray to yellowish white lesion that is round or oval.
 - *The second type* is characteristic by white, elevated, lacy, circinate lesion on the oral mucosa including the tongue. These eruption parallel those of the skin
 - *The third type* consists of a fiery red erythema of the oral mucosa, including the tongue which is seen in acute form.

- *The fourth type* of oral lesion described in psoriasis is a geographic tongue that occurs most frequently among the patients with psoriasis than without.

Diagnosis

- *Clinical diagnosis*—silvery scale appearance with positive Auspitz's signs will diagnose psoriasis.
- *Laboratory diagnosis*—intraepithelial micro-abscess formation (*Munro abscess*) is seen. There is also test tube rete pegs appearance.

Management

- *Topical agents*
 - *Emollients*—it has modest effect in terms of reducing scale.
 - *Dithranol*—it is gold standard therapy with dithranol. It inhibits proliferation when applied to psoriatic plaques.
 - *Tar*—crude tar is used which is pro-inflammatory and action same as dithranol.
 - *Calcipotriol*—it is vitamin D agonist and it reduces the thickness of plaque.
 - *Retinoid*—it diminishes the induration, scaling and redness of plaque.
 - *Corticosteroids*—use of potent topical corticosteroid on the face or hair margins should be under close and expert medical supervision.
- *Ultraviolet light*—it is mainstay of management of patient with moderate to severe psoriasis. It is used 3-7 times in a week.
- *PUVA (psoralens and ultraviolet A) therapy*—psoralens are natural photosensitizers found in number of plants. It includes clearance to greater degree than any other therapy.
- *Systemic treatment*—three main systemic agents are used, i.e. methotrexate, oral retinoid and cyclosporine.

Focal Epithelial Hyperplasia

It is also called as '*Heck disease*'. It is viral induced oral mucosal hyperplastic response characterized by multiple, more or less papillomatous like lesion.

Etiology

- *Papillomavirus*—it is possibly caused by papillomavirus type 13 and 32.
- *HIV virus*—some lesions may be seen in HIV seropositive patients.

Clinical Features

- *Age and sex predilection*—it occurs predominately in children between ages of 3 and 18 years. There is no sex predilection.

- **Site**—common sites are lip, buccal mucosa, commissures, tongue and less commonly on the gingiva, and anterior facial pillar.
- **Appearances**—it appears as multiple nodular lesions with sessile base. It can occur in cluster or in isolated crops. Some time it is present as flat, slightly raised whitish plaque on roughened surface.
- **Cobblestone appearance**—in some cases these lesions are cluster so closely the entire area appear as cobblestone or fissure appearance.
- **Sign**—they become less conspicuous when the mucosa is stretched. It is non-tender. After drying, the lesion reveals finely granular surface texture.
- **Size**—these lesions are soft having size of 1 to 5 mm in diameter with same color as adjacent mucosa.
- **Color**—they are pale to normal in color.
- **Progress**—they often appear to undergo spontaneous regression after 4 to 6 months.

Diagnosis

- **Clinical diagnosis**—sessile, pedunculated pale color clustered multiple lesions may go in favor of focal epithelial hyperplasia.
- **Laboratory diagnosis**—biopsy will show acanthosis with mild hyperparakeratosis usually present over the surface of lesion.

Differential Diagnosis

- **Condyloma acuminatum**—it is the condition with greatest similarity to focal epithelial hyperplasia. Condyloma acuminatum has got cauliflower like appearance as compared to focal epithelial hyperplasia which has got finely granular surface texture.
- **Tuberous sclerosis**—presence of cutaneous lesion.
- **Darier's disease**—there is presence of cutaneous lesion.

Management

- **Regress spontaneously**—no treatment is necessary as these are harmless lesion and it will regress spontaneously.
- **Surgical excision**—conservative surgical excision may be performed for esthetics purpose.

Oral Genodermatoses

These are also a type of keratotic lesion without increased potential for malignant transformation. They are called genodermatoses as they are genetically determined skin disorders.

White Sponge Nevus

It is oral genodermatose, which was described by Cannon in 1935, hence it is also known as 'Cannon's disease'. It is also called as 'white folded gingivostomatitis', 'congenital leukokeratosis', 'pachyderma oralis' or 'oral epithelial nevus'. It is autosomal dominant disease. It appears to represent a defect in epithelial maturation involving tonofilament formation with impaired normal desquamation of the superficial strata of cells.

Clinical Features

- **Age and sex**—it has no definite sex predilection with children most commonly affected and may be present at birth and may become intense at puberty.
- **Site**—the most common sites are cheek, palate, gingiva, floor of mouth, portion of tongue. It may be widespread and may involve entire mucosa. It can also occur on the mucous membranes of the nose, esophagus, genitalia, and rectum.
- **Appearance**—mucosa appears as thickened and folded or corrugated with soft or spongy texture and a peculiar white opalescent line. It has got soaked through, furrowed or wrinkled appearance.
- **Extent**—the lesion varies in extent from a small patch to involvement of a large area of mucosa.
- **Sign**—friction may strip superficial keratotic area leaving zone of normal looking epithelium or raw area. Ragged white area may be present which can be removed by gentle rubbing without bleeding.

Diagnosis

- **Clinical diagnosis**—diffuse thickened white folded appearance in children on buccal mucosa which doesn't disappear after stretching is typical feature of white sponge nevus.
- **Laboratory diagnosis**—biopsy shows thickened epithelium with hyperkeratosis, and acanthosis. In some cases there is extensive keratosis showing 'basket-weave' appearance.

Differential Diagnosis

- **Leukoedema**—it is only opalescent and white sponge nevus has rough granular and lethargic appearance.
- **Leukoplakia**—unusual in patients under the age of 30 years.
- **Lichen planus**—same as above.
- **Pachyonychia congenita**—presence of nail anomalies as well as skin lesions.

Management

- **No treatment**—there is no specific treatment for this disease but prognosis is very good.

Hereditary Benign Intraepithelial Dyskeratosis

It is also called as 'Witkop-Von Sallmann syndrome'. It is superficially similar to the white sponge nevus in its hereditary pattern but its clinical and microscopic features are different. It is inherited as autosomal dominant trait and there is defect in keratinization characterized by cytoplasmic accumulation of tonofilament with loss of cellular interdigitation and desmosomes.

Clinical Features

- *Age and sex*—it is most commonly seen in children with no sex predilection.
- *Sites*—it is commonly seen in eyes. Superficial gelatinous looking plaques occur on a hyperemic bulbar conjunctiva.
- *Symptoms*—there may be photophobia and blindness cause a by involvement of cornea by plaque formation and scarring.
- *Appearance*—eye is characterized by superficial, foamy, gelatinous white plaque overlying the cornea, sometimes producing temporary blindness.
- *Seasonal variation*—eye lesion shows seasonal variation tending to appear or increase in severity in the springs and disappear.

Oral Manifestation

- *Sites*—they are most commonly seen in buccal mucosa, floor of mouth, ventral and lateral surfaces of the tongue, the gingiva, and palate.
- *Appearance*—it appears generally as white, spongy, macerated lesions of the buccal mucosa with or without folds.
- *Signs*—these lesions vary from delicate, opalescent white membranous areas to a rough, shaggy mucosa.
- *Angle of mouth*—lesion frequently involves the corners of the mouth and appears as soft plaques with pinpoint elevation when the mucosa is stretched.
- *Candidial infection*—in some oral lesions, candidial infection may superimpose.

Diagnosis

- *Clinical diagnosis*—white spongy macerated lesion which is accompanied by eye lesion will diagnosed these conditions.
- *Laboratory diagnosis*—buccal mucosa exhibits thickening of the epithelium with pronounced hydropic degeneration.

Differential Diagnosis

- *White sponge nevus*—eye involvement is not present in case of white sponge nevus.

- *Lichen planus*—Wickham's striae is usually seen.
- *Mucooculocutaneous syndrome*—in this condition oral lesions are ulcerative, bullous or erythematous.

Management

- *Ophthalmologist consultation*—for eye lesion ophthalmologist consultation should be done.
- *Antifungal medication*—it is given in the condition when candidiasis superimposed on oral lesion.

Pachyonychia Congenita

It is also called as 'Jadassohn-Lewandowsky syndrome', 'Jackson-Lawler type'. It is extremely uncommon disease, inherited as an autosomal dominant characteristic with incomplete penetrance.

Clinical Features

- *Age and sex*—it usually occurs shortly after birth with no sex predilection.
- *Sites*—fingernails, toenails, palms, soles, knees, elbows, hands, and feet.
- *Nails*—there is marked thickening, increasing toward the free border with nailbed becoming filled with yellowish keratotic debris, often causing the nail to project upward at the free edge.
- *Hair and cornea*—associated sparse hair and corneal dyskeratosis producing corneal opacities have been reported.
- *Feet*—bullae formation occurs on the feet, and secondary infection of these may lead to crippling deformity.
- *Others*—thickening of the laryngeal commissures, tympanic membrane and nasal mucosa and mental retardation are also reported.

Oral Manifestation

- *Sites*—the buccal mucosa, tongue and lips.
- *Appearance*—they consist of focal or generalized white, opaque thickening of the mucosa (**Fig. 11-21**).
- *Ulcers*—there is frequent oral aphthous ulceration is seen.
- *Angular cheilitis*—in some cases inflammation of angle of mouth is seen.
- *Teeth*—natal teeth are also present.

Diagnosis

- *Clinical diagnosis*—dystrophic nail changes with thickened white appearance.
- *Laboratory diagnosis*—the mucous membrane exhibits an intracellular edema or vacuolization of the spinous cell reminiscent of white sponge nevus.



Fig. 11-21: Thickened white appearance seen on buccal mucosa in pachyonychia congenita (Courtesy Dr Pincha).

Differential Diagnosis

- *White sponge nevus*—in pachyonychia congenita there is fingernail changes in association with oral changes.
- *Lichen planus*—Wickham's striae are seen.
- *Hereditary benign intraepithelial dyskeratosis*—in these disorders there is combination of eye and oral lesion.

Management

- *No treatment*—there is no treatment for this disease.

Porokeratosis

It is also called as '*Mibelli's disease*' and it is autosomal dominant. It is characteristic by faulty keratinization of the skin followed by atrophy.

Clinical Features

- *Age and sex*—the majority of disease begins in early childhood but progression of disease is extremely slow. It appears to occur in males with greater frequency than in females.
- *Sites*—it occurs most commonly in extremities particularly in hands and feet, as well as shoulder, face and neck and the genitalia.
- *Appearance*—it consists initially of crateriform keratotic papules which gradually enlarged to form elevated plaques. In some cases there is ring-like keratotic lesion of the skin with atrophic center.
- *Size*—it ranges in size from a few millimeters to several centimeters.
- *Margins*—the plaques are surrounded by a distinct raised border of epidermal proliferation.
- *Nails*—the nails commonly become thickened and ridged. The central portion of the lesions ultimately becomes atrophic, leaving permanent scarring.

Oral Manifestation

- *Site*—it is most commonly seen on upper lip and palate.
- *Appearance*—there is numerous small slightly opalescent rings and serpiginous and hyperemic border studded over the palate.

Diagnosis

- *Clinical diagnosis*—thickened and ridge nails with opalescent palatal lesion.
- *Laboratory diagnosis*—the elevated horny margin of the lesion exhibits hyperkeratosis and acanthosis with a deep groove filled with parakeratin.

Management

- *No treatment*—there is no treatment for the disease except for removal of individual lesions.

Keratosis Follicularis

It is called as '*Darier's disease*', '*Darier-White disease*'. It is autosomal dominant trait. A lack of cohesion among the surface epithelial cells is characterized of this disease.

Clinical Features

- *Age and sex*—it is usually manifested during childhood or adolescence and has equal sex distribution.
- *Sites*—they are generally distributed above the forehead, scalp, neck, and over the shoulders.
- *Cutaneous lesion*—the cutaneous lesion appear small, firm papules (Fig. 11-22).
- *Color*—they are red when they first appear but characteristically become grayish brown or even purple later.
- *Signs*—it can ulcerate and crust over.



Fig. 11-22: Erythematous papules seen on trunk of patient in case of keratosis follicularis (Courtesy Dr Pincha).

- *Skin fold lesion*—in the skin fold the lesion tend to be coalescing and produce verrucous or vegetating macerated, foul smelling masses.
- *Nail changes*—characteristic nail changes are consisting of splintering, fissuring, longitudinal streaking and sublingual keratosis.

Oral Manifestation

- *Sites*—keratotic papule occur on oral mucosa particularly on hard and soft palate, gingiva, tongue have whitish appearance.
- *Signs*—they are multiple whitish papules which feel rough upon palpation. In some cases it has been described as rough, pebbly areas with verrucous white plaque or as having cobblestone appearance. Papules become confluent as disorder progress.

Diagnosis

- *Clinical diagnosis*—erythematous papular lesion on skin with white plaque lesion in oral cavity.
- *Laboratory diagnosis*—dyskeratosis is characterized by typical cells called *corps ronds and grains* (cell within cells).

Differential Diagnosis

- *White sponge nevus*—oral lesion of keratosis follicularis is multipapular or cobblestone appearances as compared to more diffuse or plaque-like pattern of white sponge nevus.
- *Pyostomatitis vegetans*—biopsy will afford a definite diagnosis.

Management

- *Sunscreen*—photosensitive patient should use protective sunscreen.
- *Systemic retinoid*—this is given in case of severely affected patient.

Warty Dyskeratoma

It is also called as 'isolated Darier's disease' which bears histologic similarity to keratosis follicularis but it presents as single isolated focus.

Clinical Features

- *Age and sex*—it usually occurs in older age group with male predominance.
- *Site*—the skin lesion occurs on face, scalp, neck and upper chest.
- *Appearance*—they appear as elevated nodules, umbilicated, with raised borders and varying in color from yellow or brown to gray or black (Fig. 11-23).

- *Size*—they appear as invariably single lesion varying in size from 1 to 10 mm in diameter.
- *Signs*—purulent drainage as well as bleeding occurs in some cases.



Fig. 11-23: Umbilicated nodule seen in warty dyskeratoma (Courtesy Dr Bhaskar Patle).

Oral Manifestation

- *Sites*—it is very rare and if present found most commonly on the alveolar ridge and palate.
- *Appearance*—it appears as small whitish area of the mucosa with a central depression.
- *Focal acantholytic dyskeratosis*—it is variant of warty dyskeratoma in which two or three discrete lesion arising adjacent to one another.

Diagnosis

- *Clinical diagnosis*—whitish area with central depression with skin nodule will aid in diagnosis.
- *Laboratory diagnosis*—biopsy shows central orthokeratin or parakeratin core. Epithelium shows a suprabasilar separation resulting in a cleft-like space containing acantholytic and benign dyskeratotic cells.

Differential Diagnosis

- *Keratoacanthoma*—it can attain dimension upto 1-2 cm while warty dyskeratoma are usually small. Keratoacanthoma fails to exhibit a smooth round border.
- *Molluscum contagiosum*—biopsy may require making the diagnosis.

Management

- *Conservative surgical excision*—it should be treated by surgical excision.

Pseudoxanthoma Elasticum

It is autosomal recessive and the basic defect involves the structure of elastin, making it susceptible of calcification. It is rare hereditary connective tissue disorder, characterized by generalized degeneration of the elastic fibers with a broad phenotypic expression. The clinical picture consists mainly of cutaneous, ocular, and vascular manifestations.

Clinical Features

- *Age and sex*—although widely variable, the age of onset averages 13 years with no predilection for sex.
- *Sites*—raised yellowish papules develop on areas of thickened, coarsely grained skin especially around the mouth, neck, axilla, elbows.
- *Appearance*—brownish gray streaks of the optic fundus (angioid streaks), recurrent gastrointestinal hemorrhage, weak pulse, and failing vision.
- *Cutaneous lesions*—the typical cutaneous lesions are small yellowish papules or larger coalescent plaques with an appearance similar to plucked chicken skin. More severely affected skin results in hanging, redundant folds.

Oral Manifestations

- *Sites*—mucous membranes, mainly of the inner aspect of the lower lip is affected.
- *Hound dog appearance*—skin around mouth becomes redundant, producing a 'hound dog' appearance.
- *Lip*—lower lip exhibits yellowish intramucosal nodule.

Diagnosis

- *Clinical diagnosis*—yellowish lesion seen on skin with hound dog appearance.
- *Laboratory diagnosis*—biopsy shows intramucosal nodules showing large number of thickened and twisted elastic and collagen fibers.

Management

- *No specific treatment*—at present, no specific treatment exists. The knowledge, however, of the potential complications may lead physicians to take some necessary precautions.

Hyalinosis Cutis et Mucosa Oris

It is also called as 'lipoid proteinosis' or 'Urbach-Wiethe syndrome'. It is autosomal recessive trait, characterized by subdermal and submucosal infiltration of a hyaline glycoprotein material.

Clinical Features

- *Age and sex*—it is mostly seen in young adults with no sex predilection.
- *Sites*—it is common in mucosal tissue, skin, vessel walls, larynx, and brain. It is also seen in face, eyelids and neck.
- *Symptoms*—as laryngeal mucosa and vocal cord are affected, infant may not be able to make crying sound. There is also development of hoarse voice during early childhood.
- *Brain*—there may be calcification of dorsum sellae.
- *Cutaneous lesions*—it commences as vesicles, upon healing acneform scars develop. There is also thickened furrowed appearance of skin.

Oral Manifestation

- *Sites*—most commonly affected area is lower lip. It can be seen on tongue, buccal mucosa.
- *Appearance*—affected tissue becomes infiltrated with yellowish white elevated pea sized plaque which gradually increased in size in puberty.
- *Symptoms*—restriction of oral function such as saliva flow, tooth eruption, and swallowing.
- *Angle of mouth*—radiating tissue may appear at angle of mouth.
- *Tongue*—tongue become firm and large and bound to floor of mouth. The dorsum of tongue may losses its papilla and ulcer may develop.
- *Gingiva*—diffuse hyperplastic appearing gingival infiltration is present.
- *Teeth*—there is hypodontia of maxillary lateral incisors and premolars.

Diagnosis

- *Clinical diagnosis*—aceneform scar with hypodontia, xerostomia, and delayed tooth eruption.
- *Laboratory diagnosis*—biopsy shows diffuse hyalinization with prominent hyaline perivascular cuffing. These deposits are PAS positive and show equivocal reaction for amyloid.

Differential Diagnosis

- *Focal epithelial hyperplasia, Cowden syndrome and amyloidosis*—it may show similar clinical features. A dermatological examination should be performed. Biopsy should be performed to confirm the diagnosis.

Management

- *Gingivectomy*—it is recommended when diffuse hyperplastic appearing gingival infiltration is present.

- *Debulking of tissue*—debulking of mucosal lesion may be necessary in cases when it affects the laryngeal mucosa causing difficult breathing.

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12

Oral Premalignant Lesions and Conditions

Concept of Precancer

It is applied that altered state of tissue which often, but not always, has high potential to undergo malignant transformation. Some benign lesions or conditions, for varying length of time, also generally precede oral cancer. Interestingly, these lesions or conditions share the same etiological factors with oral cancer, particularly the use of tobacco and exhibit same site and habit relationship. Many of them show high potential to become cancer and are therefore termed as pre-cancerous lesion.

It is especially important to remember that a premalignancy is not guaranteed to eventually transform into cancer, as is often but erroneously believed. Individual with oral pre-cancer has 69 times greater risk of developing oral cancer as compared to tobacco users who do not have pre-cancer.

Oral pre-cancer is distinguished into the following:

- *Pre-cancerous lesion*—it is defined as a morphologically altered tissue in which cancer is more likely to occur, than its apparently normal counter parts. For example
 - Leukoplakia
 - Erythroplakia
 - Mucosal changes associated with smoking habits
 - Carcinoma *in situ*
 - Bowen disease
 - Actinic keratosis, cheilitis and elastosis.
- *Pre-cancerous condition*—it is defined as a generalized state or condition associated with significantly increased risk for cancer development. For example
 - Oral submucous fibrosis (OSMF)
 - Syphilis
 - Sideropenic dysplasia
 - Oral lichen planus
 - Dyskeratosis congenita
 - Lupus erythematosus.

Precancerous Lesions

Leukoplakia

The term leukoplakia originates from two Greek words—*leuko* i.e. white and *plakia* i.e. patch. It is defined as any white patch on mucosa, which cannot be rubbed or scraped off and which cannot be attributed to any other diagnosable disease. The white color of results from thickened surface keratin layer.

Definition by WHO—It is a whitish patch or plaque that cannot be characterized, clinically or pathologically, as any other disease and which is not associated with any other physical or chemical causative agent except the use of tobacco.

Classification

According to clinical description

- *Homogenous*—it is completely whitish lesion.
 - *Flat*—it has smooth surface.
 - *Corrugated*—like a beach at ebbing tide.
 - *Pumice like*—with a pattern of fine lines (cristae).
 - *Wrinkled*—like dry, cracked mud surface.
- *Non-homogenous*
 - *Nodular or speckled*—characterized by white specks or nodules on erythematous base.
 - *Verrucous*—slow growing, papillary proliferations above the mucosal surface that may be heavily keratinized. Extensive lesion of this type is called as 'oral florid papillomatosis'.
 - *Ulcerated*—lesion exhibits red area at the periphery of which white patches are present.
 - *Erythroleukoplakia*—leukoplakia is present in association with erythroplakia.

According to etiology

- Tobacco induced
- Non-tobacco induced.

According to risk of future development of oral cancer

- **High risk sites**
 - Floor of mouth
 - Lateral or ventral surface of tongue
 - Soft palate
- **Low risk sites**
 - Dorsum of tongue
 - Hard palate
- **Intermediate group**
 - All other sites or oral mucosa

According to histology

- Dysplastic
- Non-dysplastic

According to extent

- Localized
- Diffuse

According to Banoczy

- *Leukoplakia simplex*—a uniform raised plaque formation, varying in size, with regular edges. It corresponds to homogenous type of leukoplakia.
- *Leukoplakia erosiva*—a lesion with slightly raised, rounded, red and/or whitish excrescence, that may be described as granules or nodules.
- *Leukoplakia verrucosa*—it is characterized by verrucous proliferation raised above the mucosal surface.

Etiopathogenesis (Fig. 12-1)**Local factors**

- **Tobacco**—it refers to dried leaves of nicotina tabacum. Tobacco is used widely in two forms: *Smokeless tobacco* (chewable tobacco and oral use of snuff) and *smoking tobacco* (cigar, cigarette, bidi and pipe).
 - **Smokeless tobacco**—when tobacco is chewed, various materials leach out of it, such as tobacco tars and resins. These are the extracts of tobacco, containing various chemical constituents such as nitrosomornicotine, nicotine, pyridine, and picoline and collidin. All these chemical constituents as well as the alkaline pH of snuff (8.2 to 9.3) act as local irritants and are related to the alterations of mucosa. Smokeless tobacco is believed to result in chemical damage that produces sub-lethal cell injury within the deeper layers of oral epithelium. This in turn induces concomitant epithelial hyperplasia. Smokeless tobacco often leads to tobacco pouch keratosis rather than true leukoplakia.
 - **Smoking tobacco**—smoking tobacco is harmful as this smoke contains polycyclic hydrocarbons, betanaphthylamine, nitrosamines, carbon monoxide, nicotine, etc. which act as source of irritation. The heat produced by smoking tobacco also plays a major

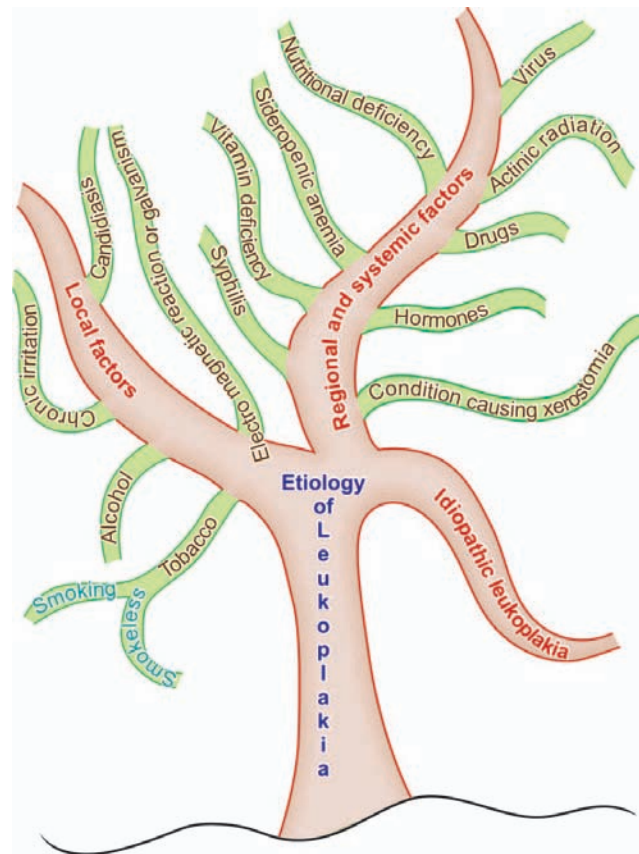


Fig. 12-1: Diagrammatic representation of etiology of leukoplakia.

role. Exposure to heat results in alteration of tissue. The initial signs of heat induced alteration of tissue are increased reddening and stippling of mucosal surface. As the use of irritant continues with the exposure to heat, minute white and red striations are formed and the tissue surface may appear slightly swollen. The striations may be caused by increased capillary proliferation and keratin formation. With continued irritation, the lesion precipitates with circumscribed configuration.

- **Alcohol**—the prevalence of leukoplakia is higher among the regular and occasional drinkers than the non-drinkers. It causes irritation and burning sensation of oral mucosa, when applied locally. Alcohol facilitates the entry of carcinogen into exposed cells and thus alters the oral epithelium and its metabolism. But most alcohol consumers use tobacco in some form or the other.
- **Sanguinaria**—this is herbal extract used in toothpaste and mouth rinse. It can cause true leukoplakia and it is more commonly seen in maxillary vestibule or on alveolar mucosa.
- **Chronic irritation**—continuous trauma or local irritation in the oral cavity is suspected as a causative agent for leukoplakia. The source of irritation or trauma may be

any viz. malocclusion, ill fitting dentures, sharp broken teeth, hot spicy food, root piece etc. The usual site to such irritation is the buccal mucosa and less often the alveolar ridge. Chronic irritation must be intense enough to induce surface epithelium to produce and retain keratin. Leukoplakia is a protective reaction of the traumatized mucosa against chronic irritation. But now-a-days it has been observed that maximum lesion due to mechanical irritation are frictional keratosis with very low precancerous potential.

- **Candidiasis**—the presence of *Candida albicans* has been reported very frequently in association with leukoplakia, more commonly with nodular type. *Candidial leukoplakia* may be associated with other local factors, such as tobacco smoking, denture wearing or occlusal friction. Tobacco smoking may result in candidal colonization because of increased keratinization, reduced salivary immunoglobulin-A concentration or depressed polymorphonuclear leukocyte function (**Fig. 12-2**).
- **Electromagnetic reaction or galvanism**—Galvanism is the generation of current due to difference in the electrical potential of two dissimilar metals. Galvanic current may arise in mouth between dissimilar, opposing or adjacent metallic restorations. Patient's complaint may range from a mere metallic taste to persistent pain, due to chronic inflammation of adjacent oral mucosa to even neuralgic pain. These mucosal changes may promote malignant transformation of leukoplakia.

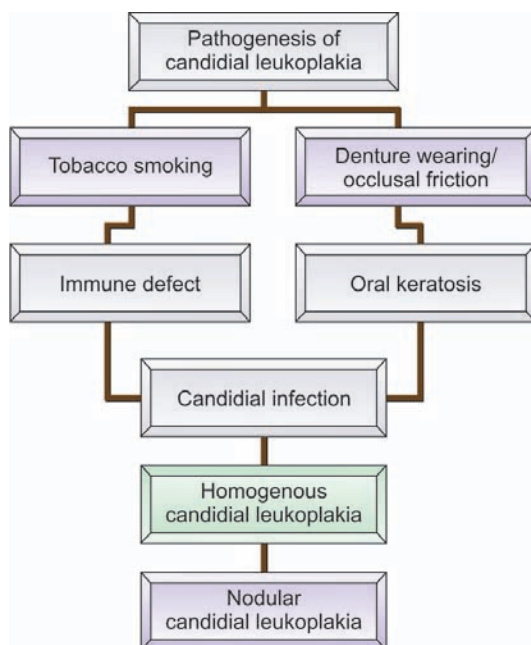


Fig. 12-2: Pathogenesis of candidial leukoplakia.

Regional and systemic factors

- **Syphilis**—it is regarded as a predisposing factor for the development of leukoplakia. White patches are often seen on tongue in tertiary syphilis. Spirochete, the causative agent for syphilis has predilection for the actively mobile tissues of tongue. These tissues are heavily involved during secondary stage of syphilis which leads to diffuse vasculitis and progresses to obliterative endarteritis, eventually resulting in a circulatory deficiency to the lingual papillae. This causes atrophy of filiform and fungiform papillae and results in bald, smooth lingual surface. Shrinkage of the lingual musculature may also occur resulting in a wrinkled surface. With the protective papillae missing, the dorsum of tongue is left extremely susceptible to oral irritation and leukoplakia frequently develops on it. Leukoplakic involvement may be minor or severe and it may be diffuse or localized. In many cases, leukoplakia is of dysplastic variety. Carcinoma of tongue frequently develops in such cases of leucic glossitis.
- **Vitamin deficiency**—deficiency of vitamin A is known to produce metaplasia and keratinization of certain epithelial structures. Hence, it may be causative factor for leukoplakia. Patients with leukoplakia show lower serum levels of vitamin A. Vitamin B complex deficiency has also been suggested as a predisposing factor. It might be related to alteration in the oxidation pattern of the epithelium, making it more susceptible to irritation.
- **Nutritional deficiency**—sideropenic anemia and other nutritional deficiency can be some predisposing factors for the occurrence of leukoplakia.
- **Xerostomia**—some conditions which can cause xerostomia may lead to leukoplakia. These conditions are salivary gland diseases, anticholinergic drugs and radiation.
- **Hormones**—it is difficult to demonstrate significance of male and female sex hormone deficiency and endocrine dysfunction in the etiology of leukoplakia.
- **Drugs**—anti-cholinergic, anti-metabolic drugs and systemically administered alcohol may predispose for the occurrence of leukoplakia.
- **Virus**—two types of viruses have been linked with leukoplakia viz. herpes simplex and human papilloma virus. A specific increase in cell mediated immunity to HSV was observed in dysplastic leukoplakia. HPV associated antigen has also been demonstrated in cases of leukoplakia. These viruses are believed to induce mucosal changes by altering the DNA and chromosomal structure of the cells and by inducing proliferation of such altered cells.
- **Idiopathic (cryptogenic) leukoplakia**—in a small proportion of cases, no underlying cause has been found. Such lesions are termed as idiopathic (cryptogenic) leukoplakia. These lesions have higher potential for malignant transformation.

Clinical Features

- *Sex and age distribution*—it occurs more commonly in older age group, i.e. 35 to 45 years and above. Males are affected more frequently than females, due to direct consequence of tobacco habit.
- *Common sites*—it can occur anywhere on the oral mucosa. Buccal mucosa (**Fig. 12-3**) and commissures (**Fig. 12-4**) are more commonly involved. Lip lesions are more common in men and tongue lesions are more common in women. The involvement of various sites depends upon the type of tobacco habit. Sometimes in edentulous patient alveolar ridge can be involved (**Fig. 12-5**).
- *Commissural leukoplakia* (**Fig. 12-6**)—the lesion is regarded as 'commissural', if it extends posteriorly from labial commissure over a distance of about 2 cm, in triangular shape. In buccal sides it involves the central zone of



Fig. 12-3: Leukoplakia occurring on the buccal mucosa.



Fig. 12-4: Leukoplakia present at commissural region.



Fig. 12-5: Leukoplakia occurring on alveolar region in edentulous patient.

buccal mucosa in the molar region and along the occlusal line. In males, commissural involvement occurs more frequently than buccal one with the latter being the commoner site in females.

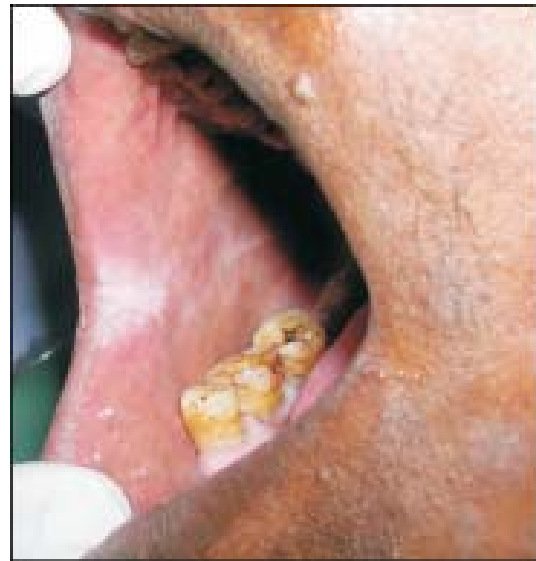


Fig. 12-6: Commissural leukoplakia presenting as triangular shaped lesion (Courtesy Dr Chole).

- *Locations in descending order of frequency of involvement*—commissures, buccal mucosa, lips, tongue, palate, and alveolar ridge, floor of mouth, soft palate and gingiva.
- *Sublingual keratosis*—it refers to leukoplakia occurring in floor of mouth and ventral surface of tongue.
- *Ebbing tide type*—some leukoplakia that occur in the floor of the mouth are referred to as 'ebbing tide' type since they appear similar to the undulations left on the sand by the ebbing tide. It occurs due to loose binding and consequent movement of the mucosa in the floor of mouth.

- **Extent**—the extent of involvement may vary from small, well-localized, irregular patches to diffused lesions involving considerable portion of oral mucosa (**Fig. 12-7**). Multiple areas of involvement are not uncommon.



Fig. 12-7: In this leukoplakia, considerable portion of oral mucosa is involved.

- **Color**—lesion may be white or yellowish white (**Fig. 12-8**), but with heavy use of tobacco lesion it may assume brownish color.
- **Surface**—the surface of the lesion is often finely wrinkled or shriveled in appearance and may feel rough on palpation.



Fig. 12-8: Yellowish white color of leukoplakia seen on buccal mucosa. Surface seen as wrinkled (Courtesy Dr Suwas Darvekar).

- **Symptoms**—some patients may report a feeling of increased thickness of mucosa. Those with ulcerated and nodular type may complain of burning sensation. Enlarged cervical lymph nodes may be a single occurrence of metastasis.

Clinical Types

- **Pre-leukoplakia or mild or thin leukoplakia**—a different entity termed as 'pre-leukoplakia' has been distin-

guished. It is a low grade or very mild reaction of the mucosa appearing as grayish white but not completely white lesion with slight lobular pattern. Their margin can be well demarcated or it blends with the adjacent normal mucosa. These lesions are soft, translucent and thin (**Fig. 12-9**).



Fig. 12-9: Mild or thin type of leukoplakia seen in buccal mucosa in posterior region.

- **Homogenous or thick leukoplakia**—it is also called as *leukoplakia simplex*. It accounts for 84% of cases. Leukoplakia seen amongst clay pipe smokers and betel quid chewers are generally of homogenous type. They have no red component.
 - **Appearance**—usually, localized lesions of extensive white patches present a relatively consistent pattern throughout (**Figs 12.10A and B**). However, sometimes the surface lesion may be described as corrugated, with pattern of fine lines, or wrinkled or papillomatous surface.
 - **Size and margin**—it is characterized by raised plaque formation consisting of single or group of plaques varying in size with irregular edges.
 - **Color**—they are usually white in color but may be yellowish white or yellow.
- **Ulcerated leukoplakia**—it occurs in 13% of cases. It is characterized by red area, which at times exhibit yellowish areas of fibrin, giving the appearance of ulceration. White patches are present at the periphery of the lesion (**Fig. 12-11**). Sometimes, it is associated with pigmentation of varying intensity, usually on the periphery of the lesion. Heat produced during smoking also contributes to the occurrence of pigmentation.
- **Nodular leukoplakia**—it is also called as '*leukoplakia erosiva*' or '*speckled leukoplakia*'. It is a mixed red white lesion in which small keratotic nodules are scattered over an atrophic patch of oral mucosa. Nodules may be pinhead sized or even larger (**Fig. 12-12**). It has got a high malignant potential.



Figs 12-10A and B: Homogenous leukoplakia showing thick white patches on buccal mucosa (Courtesy Dr Parate).



Fig. 12-11: Ulcerated type of leukoplakia showing white patches at periphery in the buccal vestibule. Patient is also having leukoplakia at the commissural region extending into buccal mucosa.

- **Verrucous leukoplakia or verruciform leukoplakia**—it is also called as '*leukoplakia verrucosa*'. It is characterized by verrucous proliferation above the mucosal surface. These lesions demonstrate sharp and blunt projection (Fig. 12-13). These projection are heavily keratinized.



Fig. 12-12: Nodular leukoplakia showing pinhead size nodule in retromolar area.



Fig. 12-13: Verrucous leukoplakia showing exophytic lesion present on the tongue (Courtesy Dr Parate).

They may be accompanied by homogenous leukoplakia on other oral mucosal surfaces.

- **Proliferative verrucous leukoplakia (PVL)**—verrucous leukoplakia can become more exophytic with development of multiple keratotic plaques with roughened surface projection (Fig. 12-14). PVL has got strong female predilection and is present in patient who don't use tobacco. It can spread and involve adjacent mucosa. As the disease progress it transforms into a lesion that is clinically and microscopically identical to verrucous carcinoma or squamous cell carcinoma.
- **Erythroleukoplakia**—in some lesion of leukoplakia red component is present. This intermixed lesion is called as erythroleukoplakia

Staging of Leukoplakia

According to size, clinical aspect and pathological features

- **Size**—it is denoted by L
 - L_1 —size is less than 2 cm.
 - L_2 —size is in the range of 2-4 cm.



Fig. 12-14: Proliferative verrucous leukoplakia showing multiple keratotic clefts in anterior region.

- L_3 —size is more than 4 cm.
- L_x —size is not specified.
- **Clinical aspect**—it is denoted by C
 - C_1 —homogenous
 - C_2 —non-homogenous
 - C_x —not specified
- **Pathological features**—it is denoted by P.
 - P_1 —no dysplasia
 - P_2 —mild dysplasia
 - P_3 —moderate dysplasia
 - P_4 —severe dysplasia
 - P_x —not specified
- **Site**—site of leukoplakia
 - S_1 —all sites excluding floor of mouth, tongue
 - S_2 —floor of mouth and tongue
 - S_3 —not specified.

Sharp staging

- **Stage I**—Earliest lesion—non-palpable, faintly translucent, white discoloration.
- **Stage II**—Localized or diffused slightly elevated plaque of irregular outline. It is opaque white and may have fine granular texture.
- **Stage III**—Thickened white lesion showing induration and fissuring.

Diagnosis

- **Clinical diagnosis**—clinically any white patch with history of tobacco chewing which cannot be rub off is the diagnostic indicator for leukoplakia.
- **Laboratory diagnosis**—in biopsy you can see *hyperorthokeratosis, hyperparakeratosis, acanthosis of epithelium, epithelial dysplasia, liquefaction degeneration basal cell hyperplasia*. Scanning electron microscopy will show epithelial dysplastic changes.

Malignant Potential

The term malignant transformation is used to denote development of oral cancer from pre-existing leukoplakia. Malignant transformation occurs in 0.3% to 10% of cases.

It is higher in women (6%) than men (3.9%), due to involvement of endogenous factors. Leukoplakia associated with chewing habit of tobacco shows higher rate of transformation as compared to others. In buccal mucosa and commissure region 1.8% malignant transformation occurs. In lip and tongue region 16% to 38.8% malignant transformation occurs. Nodular dysplasia has higher risk of malignant transformation than other clinical types. Idiopathic leukoplakia and candida associated leukoplakia also come under high risk.

If following features are present there is a high risk of malignant transformation:

- Persistence of lesion for several years.
- Female patient
- Lesion situated on the margins, base of tongue and floor of mouth
- Erosive lesions.
- Combination of above factors.

Differential Diagnosis

- **Lichen planus**—distinguished by the often occurrence of multiple lesions and presence of 'Wickham's striae'.
- **Chemical burn**—history is important in chemical burn.
- **Syphilitic mucus patches**—other features like split papule or condyloma latum may be present.
- **White sponge nevus**—occurs soon after birth or at least by puberty and is widely distributed over the oral mucus membrane. Familial pattern is seen, in contrast to leukoplakia which occurs over 40 years of age and not so disseminated throughout the oral cavity.
- **Discoid lupus erythematosus**—central atrophic area with small white dot and slightly elevated border zone or radiating white striae.
- **Psoriasis**—Auspitz's sign is positive and skin lesions are also present.
- **Leukoedema**—classically occurs on buccal mucosa covering most of the oral surface of cheek and extending onto labial mucosa. Faint milky appearance with folded and wrinkled pattern as compared definite whiteness of leukoplakia.
- **Hairy leukoplakia**—corrugated leukoplakic lesion occurring on lateral and ventral surface of tongue in patients with AIDS or ARC (AIDS related complex).
- **Verruca vulgaris**—commonly occurs in the oral cavity as small, raised white lesion, more than 0.5 cm in diameter as compared to verrucous leukoplakia which is larger and surrounded by inflamed mucosa.

- *Verrucous carcinoma*—lesions are elevated (exophytic).
- *Cheek biting lesion*—careful history elicits the cause and promotes proper diagnosis.
- *Electrogalvanic white lesion*—disappears when different metal restorations are replaced with composite restoration or when teeth are extracted.

Management

Elimination of etiological factors

- *Prohibition of smoking*—patient should be asked to cease smoking immediately. Many cases of leukoplakia get regress after smoking is ceased.
- *Removal of chronic irritant*—dentist should remove sharp, broken teeth.
- *Elimination of other etiological factors*—other etiological factors like syphilis, alcohol, dissimilar metal restoration etc. should be eliminated.

Conservative treatment

- *Vitamin therapy*—it has a protective effect on the epithelium. Daily requirement is 4000 IU. It is given orally, parentally or topically. Therapeutic dose—75000-300000 IU for 3 months. Vitamin A may be used topically after painting the lesion with podophyllin solution (it inhibit mitosis).
- *Vitamin A + vitamin E*—this therapy is given to inhibit metabolic degradation.
- *13-cis-retinoic acid*—It is a synthetic analogue of vitamin A; usually given in high doses of 1.5 to 2 mg/kg body weight for 3 months. If relapse occurs then low doses of 13-cis-retinoic acid 0.5 mg/kg body weight are given for 9 months. It may be given with β carotene (this drug may produce a variety of toxic effects which include facial erythema, dryness, peeling of skin, conjunctivitis and hypertriglyceridemia).
- *Antioxidant therapy*—considerable data shows that β -carotene supplementation can be beneficial for treatment of oral leukoplakia.
- *Vitamin A palmitate*—Short-term treatment with vitamin A palmitate along with aromatic retinoid of all trans -B-A vitamin acid plus, B-cis-B-A vitamin acid may show healing and improvement.
- *Nystatin therapy*—It is given in candidial leukoplakia. 500,000 IU twice daily plus 20% borax glycerol or 1% gentian violet or mouth rinses with chlorogen solution.
- *Vitamin B complex*—it is given as supplement in cases of commissural and lingual lesion.
- *Antimycotic preparation*—the antimycotic preparation can hasten and pimafucin have also been effective.
- *Panthenol*—panthenol lingual tablet and oral spray may be used against glossitis and glossodynia, in case of tongue lesion.
- *Estrogen*—in some cases, administration of estrogen can be helpful.

Surgical management

To give proper treatment, microscopic examination is necessary for which biopsy is taken. The most meaningful sites for taking biopsy specimen are the areas that display greater surface irregularities such as cracks and fissures and those associated with erythematous areas.

- *Conventional surgery*—you should make an incision around the lesion including safe margins. The incision should be deep and wide. Affected area is then undermined and dissected from the underlying tissue. Sliding mucosal flap is prepared for covering the wound. Fine iris scissors and skin hook should be used to decrease trauma. Fairly extensive undermining of the mucosal flap is necessary so that when it is advanced into position, there will be minimum amount of tension. Excessive tension will restrict circulation and can result in necrosis. After proper mobilization of the mucosal flap, it is advanced and multiple interrupted black silk sutures are used. Approximate the free edges. Postoperative application of ice bags to the site is advised to minimize bleeding and swelling.
- *Cryosurgery*—tissue is exposed to extreme cold to produce irreversible cell damage. Cell death occurs at -20 degree Celsius. Disc type probe (cryo probe) refrigerated by liquid nitrogen or pressurized nitrogen oxide is used. Incorporated in the probe is re-warming device, thus tissue can be rapidly frozen and thawed as required. The probe is applied to the surface lesion, which is moistened by water-soluble jelly. It produces very low temperature in tissue. First freeze for 1 minute, followed by 5 minute thaw. 1 minute freeze and then administered. Freezing produces white area of necrosis. The process is repeated two to three times to achieve maximum destruction. Freezing induces crystal formation within the cell and intracellular spaces. When cooling rates are rapid, intracellular ice crystal may rupture cell membrane and cell death can occur. Slower cooling produces large ice crystals in the space between cells. As the crystals grow, water is removed from neighboring cells, leading to an increased concentration of the electrolytes. These soon reach toxic levels initiating cell death by osmotic shock when thawing commences. Cell is also killed by change in microvasculature of the part being exposed to cryosurgery.
- *Fulguration (electrocautery and electrosurgery)*—It is a technique in which there is destruction of tissues by high voltage electric current and the action is controlled by movable electrode. Advantages of this are its ability to coagulate lesion and provide easy control of hemorrhage. Disadvantages are tasteless (foul) odor, hazards of explosion, need for profound local anesthesia and sometimes general anesthesia, slow healing process, pain and scarring.

LASER

LASER stands for light amplification of stimulated emission of radiation. CO₂ lasers are most commonly used in oral lesions due to their great affinity for any tissue with high water content and their minimum penetration depth, i.e. 0.2 to 0.3 mm in oral tissue. CO₂ laser contain CO₂, nitrogen and helium gases

- **Biopsy**—in performing incisional or excisional biopsy in cases of leukoplakia, CO₂ laser is used as precise cutting tool. Laser is placed in a cutting or focused mode held perpendicular to the tissue. A predetermined surgical outline is followed by means of a tissue pickup; a border of the outline is raised and the lesion is then undermined with traction and counter traction would be done with scalpel.
- **Laser peel**—it is usually used to remove the lesion that involves relatively large surface area. Beam is highly defocussed and kept distant from the tissue. Initially, no effect will be seen on the tissue plane Beam is gradually brought closer into focus, but is still in defocussed mode until tissue takes white appearance and begins to blister. Blistering occurs at the basement membrane of the tissue. This whitening technique is extended over the rest of the lesion to be peeled. This white area is then grafted with tissue forceps or hemostat and it is 'laser peeled' away to expose the underlying tissue or vitalization.

- **Ablation**—it is the method of painting away a lesion or tissue removed by laser. It is a non-contact surgical application in which tissue is simply vaporized. It is particularly applied to larger areas of leukoplakia, when surgical excision will create major problems with morbidity and reconstruction. Surface vaporization to a depth of approximately 0.5 mm will normally result in good secondary re-epithelization with normal mucosa. During surface ablation, carbonized layer should be removed before deeper layer is ablated.

Miscellaneous

- **Radiation therapy**—It is utilized only in neoplastic tissue. In some cases, it has been tried, but late observation has failed to show complete success.
- **Topical chemotherapy**—topical application of anticancer chemotherapeutic agent such as bleomycin and human fibroblast interferon have been used with success in limited cases of leukoplakia.

Important Guidelines for Treatment of Leukoplakia (Fig. 12-15)

- Biopsy should be done.
- Leukoplakia that do not heal in 2 to 3 weeks after elimination of demonstrable etiological factors, require conservative and surgical treatment.

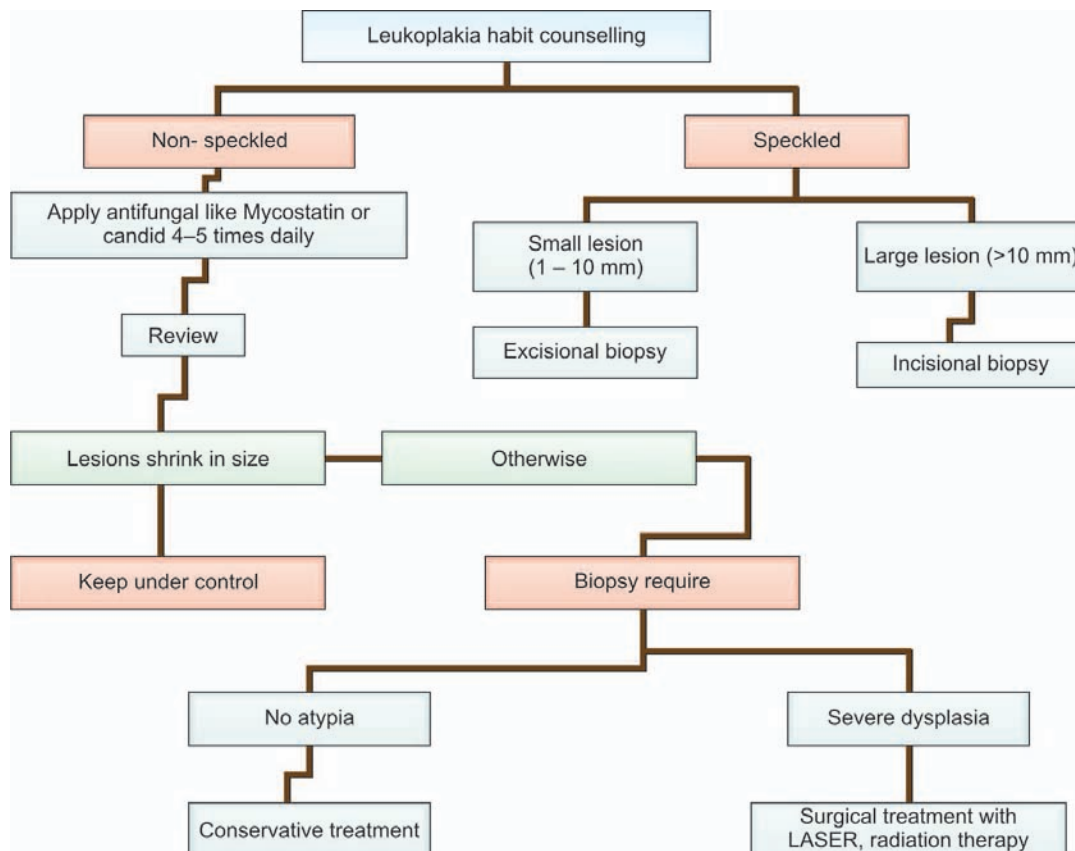


Fig. 12-15: Guidelines for the management of leukoplakia in dental office.

- Conservative treatment is applied to a large incipient lesion with keratinization and large verrucous lesions, in order to reduce their size before surgical intervention.
- Surgical treatment is applied if conservative treatment fails to produce complete healing in 3 months at latest.
- Because of higher risk of nodular and verrucous leukoplakia to become malignant, excision of these lesions with adequate follow-up is necessary.
- To prevent malignant changes, all patients reported with leukoplakia are summoned for re-examination at least twice a year.
- Re-biopsy after 5 to 6 months.
- 6 to 10% of leukoplakia with evidence of dysplasia progress to malignancy.
- Recurrence will often follow excision.

Erythroplakia

It is also called as *erythroplasia of Queyrat*, *erythroplasia*. Erythroplakia is a persistent velvety red patch. Reddish color results from absence of surface keratin layer and due to presence of connective tissue papillae containing enlarged capillaries projected close to the surface.

Evolution of the Disease

The first mention of a non-hemorrhagic red patch of an upper aerodigestive tract site was in a vocal cord lesion reported in 1852. The first truly descriptive paper of erythroplakia, however, was a 1911 report by Queyrat, of a red macule on the glans penis of a syphilitic patient. It was Queyrat who coined the term "*erythroplasia*."

About the same time, Rubin used the term "*incipient carcinoma*" to describe the microscopic features of erythroplakia, most common histopathologic diagnosis as carcinoma in situ, based on his experience with lesions of uterine cervix.

Definition

- It is applied to any area of reddened, velvety-textured mucosa that cannot be identified on the basis of clinical and histopathologic examination as being caused by inflammation or any other disease process.
- A chronic red mucosal macule which cannot be given any other specific diagnostic name and cannot be attributed to traumatic, vascular or inflammatory causes.

Classification

- *Homogenous*—these commonly occur on buccal mucosa with well demarcated margin.
- *Erythroleukoplakia*—erythroplakia interspersed with patches of leukoplakia.
- *Granular or speckled*—these are elevated lesions.

Etiology

- *Idiopathic*—in most of the cases cause for erythroplakia is not found.
- *Alcohol and smoking*—these can act as predisposing factors for erythroplakia.
- *Candida infection*—a secondary infection or super infection with candidiasis may be associated with dysplastic oral mucosal cells. *Candida albicans* has often been demonstrated in erythroleukoplakia lesions and the red component of these lesions (often the white component as well) diminishes or disappears after antifungal therapy, in at least some cases.

Clinical Features

- *Age and sex*—male predilection and most common in 6th and 7th decade of life.
- *Sites*—erythroplakia occurs on all mucosal surfaces of the head and neck area. Half of all cases, however, are found on the vermilion or intraoral surfaces, with the rest being evenly divided between the larynx and the pharynx. Vermilion lesions are relatively common and are most often seen on the lower lip. Intraorally, the lateral and ventral tongue, floor of mouth, buccal vestibule and soft palate are most frequently involved.
- *Symptoms*—erythroplakia, as the name obviously implies, is asymptomatic.
- *Appearance*—it is nonelevated, red macule or patch on an epithelial surface. The exact cause of the red appearance is unknown, but may be related to an increase in the number of underlying blood vessels through which the blood flows, which in turn may be secondary to localized inflammatory or immunological responses caused by the dysplastic, i.e. 'foreign,' epithelial cells. In some cases, the color may result from a lack of surface keratin or extreme thinness of the epithelium.
- *Extent*—unlike leukoplakia, erythroplakia is seldom multiple and seldom covers extensive areas of mouth. Also unlike leukoplakia, erythroplakia seems seldom to expand laterally after initial diagnosis, although this may be an artifactual feature because most lesions are completely removed or destroyed immediately after formal diagnosis.
- *Homogenous form*—homogenous form appears as a bright red, soft, velvety lesion with straight or scalloped, well-demarcated margins (**Fig. 12-16**). It is often quite extensive in size. Regardless of the cause of the color change, the typical lesion is less than 1.5 cm in greatest diameter and half are less than 1.0 cm, but lesions larger than 4 cm have also been seen. It usually quite sharply demarcated from the surrounding pink mucosa and its surface is typically smooth and regular in coloration.



Fig. 12-16: Homogenous type of erythroplakia with bright red lesion with well demarcated margin seen in buccal vestibule.

- *Granular or speckled form*—these are soft, red lesions that are slightly elevated with irregular outlines and granular or finely nodular surface speckled with tiny white plaques (**Fig. 12-17**).
- *Smooth erythroplakia* is soft to palpation and has often been described as having a velvety feel. The pebbled lesions tend to be somewhat firm, but erythroplakia never actually becomes hard or indurated, until an invasive carcinoma develops within it.
- *Erythroleukoplakia*—it is quite common to see erythroplakia admixed with or adjacent to leukoplakia in mouth. In such lesions, the red areas are the sites most likely to contain or to develop dysplastic cells and should therefore be the sites most readily biopsied and most carefully examined clinically. Erythroplakia interspersed with patches of leukoplakia in which erythematous areas are irregular and often not as bright as homogenous form, are most frequently seen on tongue and floor of mouth. The borders may be well circumscribed or blend impercibly with surrounding oral mucosa.



Fig. 12-17: Granular form of erythroplakia showing elevated irregular outline (Courtesy Dr Suwas Darvekar).

Diagnosis

- *Clinical diagnosis*—red well demarcated path with no sign of infection and inflammation give rise to diagnosis of erythroplakia.
- *Toluidine blue test*—Differentiation of erythroplakia with malignant changes and early squamous cell carcinoma, from the benign inflammatory lesions of oral mucosa is enhanced by use of 1% toluidine blue test. The solution is applied locally by swab or oral rinse. Malignant type retains it, owing to increased nuclear DNA content of tumor cells.
- *Laboratory diagnosis*—biopsy exhibits epithelial changes ranging from mild dysplasia to carcinoma in situ and even invasive carcinoma.

Differential Diagnosis

- *Candidiasis*—lesion can be rubbed off and it is commonly seen on the tongue.
- *Denture stomatitis*—unusually site is the palate or any denture bearing area.
- *Tuberculosis*—tuberculous ulcers are present which have rolled margins.
- *Histoplasmosis*—it is more common in farmers and present as a single ulcer.
- *Area of mechanical irritation*—cause can be identified.
- *Macular hemangioma*—lesions blanch on pressure.
- *Talangiectasia*—characteristic appearance seen on soft palate.
- *Traumatic lesion*—cause can be identified.

Management

- *Removal of cause*—elimination of a suspected irritant should be carried out.
- *Incisional biopsy*—it is always preferred method for establishing a microscopic diagnosis of suspicious intraoral lesions. Since erythroplakia is so closely correlated with severe dysplasia, carcinoma in situ and invasive carcinoma, incisional biopsy is especially indicated. Excisional biopsy of a potential malignancy may result in under treatment and violation of surgical oncologic principles.
- *Surgical stripping*—the definitive treatment of erythroplastic lesions remains controversial. A conservative surgical procedure such as mucosal stripping is often performed, with minimal damage to the deeper connective tissues. This has the distinct advantage of preserving tissues for microscopic evaluation of potential regions of invasion.
- *Destructive technique*—destructive techniques such as laser ablation, electrocoagulation and cryotherapy have also proved to be effective.

- **Clinical follow-up**—the key to therapy in this disease is extended clinical follow-up. Patients should be examined every 3 months for the first postoperative year and every 6 months for an additional 4 years. After that, annual reevaluation with a thorough head and neck examination is advisable.

Carcinoma in situ

It is also called as '*intraepithelial carcinoma*'. Severe dysplastic changes in a white lesion indicate considerable risk of development of cancer (**Fig. 12-18**). The more severe grade of dysplasia merges with the condition known as carcinoma in situ. It is more common on skin but can also occur on mucous membrane.

Clinical Features

- **Age and sex**—male predilection and occurs more commonly in elderly persons.
- **Sites**—common sites are floor of mouth, tongue and lips.
- **Appearance**—appearance of the lesion may be like leukoplakia and erythroplakia. It may be a combination of leukoplakia and erythroplakia, ulcerated lesion, ulcerated and white lesion, red and ulcerated lesion or may be non-specific.

Diagnosis

- **Clinical diagnosis**—clinically one cannot diagnose carcinoma *in situ*. It may appear like leukoplakia, erythroplakia or both.
- **Laboratory diagnosis**—in biopsy, keratin may or may not be present in/on the surface of lesion; but if present, is more apt to be parakeratin, rather than orthokeratin. Loss of orientation of cells and their polarity (**Fig. 12-18**). Sharp line of division between normal and altered epithelium extending from the surface, down to the connective tissue rather than blending of the epithelium. An increase in nuclear/cytoplasmic ratio and nuclear hyperchromatism are sometimes seen.



Fig. 12-18: Carcinoma *in situ* showing dysplastic changes.

Management

- **Surgical removal**—lesion may be surgically excised, cauterized and even exposed to solid carbon dioxide.

Actinic Keratosis

Actinic keratosis is a cutaneous premalignant lesion. Similarly actinic cheilitis which is associated with lower lip is described in Chapter 23: Diseases of Lip.

Etiology

- **Ultraviolet radiation**—this radiation can produce mutation in p53 suppressor gene. This will cause alteration in skin.

Clinical Features

- **Age and sex distribution**—it is more common in older age group. It is more commonly seen in men.
- **Site**—it is more commonly seen on face, neck, dorsum of hands, the forearms and scalp of bald headed men.
- **Appearance**—they are irregularly scaly plaques. Background is erythematous.
- **Color**—they vary in color from normal white, gray, brown.
- **Size**—the size of lesion is usually less than 7 mm in diameter.
- **Sandpaper texture**—palpation of the lesion shows/feels roughened texture which resembles as sandpaper.
- **Keratin horn**—in some cases, lesion may produce so much keratin that it appears as keratin horn from central area.

Management

- **Cryotherapy**—it is done with the help of liquid nitrogen.
- **Other therapy**—other therapy like topical application of 5-fluorouracil, curettage, electrodesiccation or surgical excision.

Oral Lesions Associated with Use of Tobacco

Stomatitis Nicotina

It is also called as '*smoker's palate*', '*stomatitis nicotina palati*'. It refers to a specific white lesion that develops on the hard and soft palate in heavy cigarette, pipe, and cigar smokers. In many cultures, hand rolled cigarettes and cigars are smoked with the burning end held within the mouth. This habit is called as '*reverse smoking*' and the lesion associated with it is called as '*reverse smoker palate*'. The reason for

occurrence of the lesion is due to heat rather than an effect of tobacco (**Fig. 12-19**).



Fig. 12-19: Well defined reddened area seen in palate of patient who is having habit of reverse chutta smoking (Courtesy Dr Chole).

Types (Clinical)

- *Mild*—consisting of red, dot like opening on blanched area.
- *Moderate*—characterized by well defined elevation with central umbilication.
- *Severe*—marked by papules of 5 mm or more with umbilication of 2-3 mm.

Palatal Change in Reverse Smoking

- *Keratosis*—diffuse whitening of the entire palatal mucosa.
- *Excrescences*—1-3 mm elevated nodules, often with central red dots corresponding to the opening of palatal mucous glands.
- *Patches*—well defined, elevated white plaques, which could qualify for the clinical term leukoplakia.
- *Red areas*—well defined reddening of the palatal mucosa.
- *Ulcerated area*—crater-like areas covered by fibrin.
- *Non-pigmented areas*—area of palatal mucosa which is devoid of pigmentation.

Clinical Feature

- *Age and sex distribution*—it is usually seen in men who are pipe smokers. It is common in middle age and elderly adults.
- *Site*—most commonly affected site is palate. The lesion is well developed and prominent on keratinized hard palate. It is restricted to the area which is exposed to heavy cigarette smoke.

- *Onset*—initially there is redness and inflammation of the palate.
- *Appearance*—In the early stages, mucosa is reddened. It subsequently becomes grayish white (**Fig. 12-20**), thickened and fissured. Fissures and cracks may appear producing a wrinkled, irregular surface. In some cases, there may be papular or multinodular appearance. Papules do not coalesce and are separated from one another by intervening normal appearing mucosa. Tonsillar pillars are usually erythematous.



Fig. 12-20: Grayish white appearance of palate seen in smoker's palate.

- *Dried mud appearance*—in some cases, palatal keratin becomes so thickened that it impart fissure or dried mud appearance (**Fig. 12-21**).

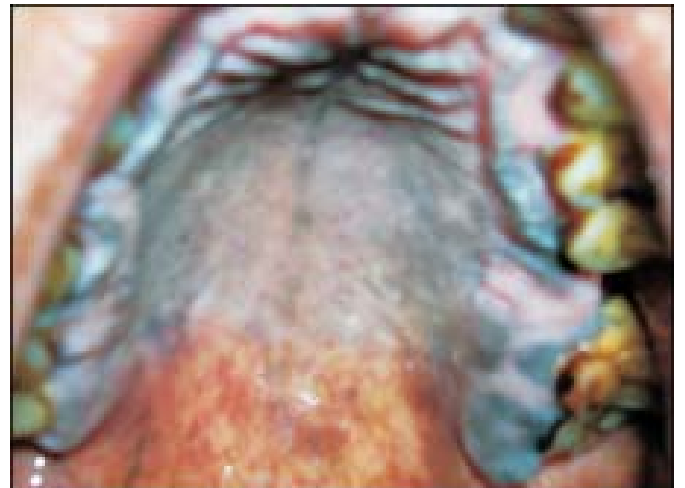


Fig. 12-21: Dried mud appearance seen in stomatitis nicotina.

- *Salivary gland opening*—discoloration is homogenous with the exceptions of numerous erythematous spot (**Fig. 12-22**). It represent focal thickening surrounding the orifice of the salivary gland which appears as white umbilicated nodule with red center that may be stain brown by deposit of tar.



Fig. 12-22: Salivary gland opening seen as numerous erythematous spots in palate.

Diagnosis

- *Clinical diagnosis*—history of cigar or pipe smoking and reverse smoking with generalized lesion of palate is key for the diagnosis of stomatitis nicotina.
- *Laboratory diagnosis*—in biopsy epithelium shows acanthosis and hyperkeratosis. Epithelium lining of minor salivary gland often shows squamous cell metaplasia and hyperplasia.

Differential Diagnosis

- *Papillary hyperplasia*—the lesion displays cobblestone appearance and in stomatitis nicotina, there is red center located on the palate of pipe or cigar smokers. The papules of the papillary hyperplasia are focal.
- *Darier's disease*—they appear diffusely on palate in cobblestone pattern.
- *Focal epithelial hyperplasia*—it is not common on palate and they are not erythematous.
- *Cowden syndrome*—these are multiple papillary nodules commonly seen on gingiva.

Management

- *Stoppage of habit*—it is completely reversible, once the habit is discontinued. The lesions usually resolve within 2 weeks of cessation of smoking.
- *Biopsy*—biopsy of nicotine stomatitis is rarely indicated. But biopsy should be performed on any white lesion of the palatal mucosa that persists after 1 month of discontinuation of smoking habit.

Snuff Dipper Lesion

It is also called as *snuff pouch*, *tobacco pouch keratosis*, *spit tobacco keratosis*, *smokeless tobacco keratosis*. The habit of chewing tobacco is called as *smokeless tobacco* or *spit tobacco* use.

Etiopathogenesis

- *Nicotine*—the compound N-nitroso-nor-nicotine (NNN), which is derived partly from bacterial action on nicotine during the curing process, is contributed by the action of salivary nitrites, when tobacco is held in the mouth; occurs in greater concentration in snuff tobacco.

Clinical Features

- *Location*—it occurs in mucosal surface, where snuff is habitually held.
- *Gingiva and periodontal tissue*—there is painless loss of gingival and periodontal tissue in the area of tobacco contact.
- *Teeth*—there is extrinsic stain present on the teeth. There is also cervical erosion of teeth with more prevalence of dental caries.
- *Smokeless tobacco keratosis*—it is white plaque present in the mucosa where chewing tobacco is kept. It is thin, gray or gray-white translucent lesion (Fig. 12-23). Margin of the lesion blends gradually into surrounding mucosa.



Fig. 12-23: Grayish lesion seen in buccal vestibule of patient where he uses to keep tobacco.

The appearance of lesion depends upon hours of daily use and use of different tobacco leaves.

- *Snuff pouch, tobacco pouch*—mucosa is soft velvety touch feel on palpation and stretching of mucosa reveal distinct 'pouch'. Stretched mucosa appears fissured and ripped in sand on a beach after an ebbing tide.
- *Malignant transformation*—verruccous carcinoma has been reported to occur from snuff dipper lesion. This is also called as snuff dipper cancer.

Management

- *Stoppage of habit*—maximum lesion is regress following the caseation of habit.
- *Biopsy*—any lesion which remains after 6 months quitting the habit should be send for biopsy.

Cigarette Smoker's Lip Lesion

They are generally flat or slightly elevated nodular white lesion on one or both lips, corresponding to the site at which the cigarette is held and apparently smoked down to an extremely short length.

There is increased redness and stippling of lip in localized area. Margin has elliptical, circular or irregular borders. Color is pale to white (Figs 12-24A and B) and is slightly elevated with nodular or papillary shape.

Pre-malignant Conditions

Lichen Planus

Erasmus Wilson described it in 1869. The term lichen planus is derived as lesion they look like lichen on the rock and planus is for flat. Various mucosal surfaces may be involved, either independently or concurrently, with cutaneous involvement or serially. Oral mucosa is frequently involved. It is a probable pre-cancerous condition. Lichen planus is a common inflammatory disease of the skin presenting with characteristic violaceous, polygonal, pruritic papules. The disease may also affect the mucosa, hair and nails.

It is relatively common dermatological disorder occurring on skin and oral mucous membrane and refers to lace-like pattern produced by symbiotic algae and fungal colonies on the surface of rocks in nature (lichens).

Prevalence of lichen planus in general population is about 0.9% to 1.2% and prevalence of oral lichen planus is reported between 0.1% and 2.2%.

Etiology

- *Cell mediated immune response*—it is a cell mediated immune response associated with lymphocyte-epidermal interactions, resulting in degeneration of basal cell layer. This occurs due to alternation of



Fig. 12-24: Cigarette smoker's lip lesion. (A) Lesion on the lower lip. (B) Lesion on the lower lip.

keratinocytes as a result of unknown events resulting in antigenic alternation of these cells thereby stimulating immunological reaction. Another hypothesis is that it is a primary immunologic reaction causing alternation and degeneration of keratinocytes. Cell mediated immune response may be caused by various mononuclear cells i.e. langerhans cell, macrophages predominantly T lymphocytes, lymphoblast cells, B lymphocytes and mast cells. These cells infiltrate the upper part of lamina propria of submucosa. The macrophages are mostly mature, which probably have functional role with mononuclear cells suggesting of cell-to-cell co-operation.

- *Recent hypothesis of pathogenesis of lichen planus.* In a genetically predisposed individual, haptens (certain drug or dental material), conventional antigen or super-antigen of oral microbial origin can induce cell mediated immune response resulting in sub-epithelial T cell infiltration of the site in oral mucosa with cytokine generation HSP-60 and C 1/10 expression by basal keratinocytes. If individual is not predisposed to react to HSP-60, then non-specific mucositis occur. If the

individual has genetic predisposition, it results in autoimmune reaction → activation of cytotoxic T cell → destruction of basal keratinocytes → oral lichen planus.

- *Auto-immunity*—the activated T lymphocytes also secrete gamma interferons which induce keratinocytes to produce HLA-DR and increase their rate of differentiation with formation of thickened surface. Antigenic information is transferred from 'Langerhans' cells to lymphocytes, when there is mutual expression of HLA-DR. Lymphocytes normally are attracted towards HLA-DR expressing keratinocytes and may contact the epithelial cell. During this contact, inappropriate epithelial antigenic information may be passed to lymphocytes due to HLA-DR linkage. With this mechanism, self antigen may be recognized as foreign bodies, leading to destruction of basal cells, resulting in an autoimmune response. There are numerous studies which show immune deposits in lichen planus affected tissues but it is not specific.
- *Immunodeficiency*—there has been report of decreased serum levels of IgG, IgA, or IgM in lichen planus and the possibility of it as a manifestation of immunodeficiency has been raised. But at the same time, reports of normal concentrations of IgA and IgM are found; therefore the role of immunodeficiency is questionable.
- *Genetic factors*—cases of lichen planus are reported in families, twins and husband and wife. Clinically, familial lichen planus is somewhat unusual as it appears to affect young patients, is severe, often extensive, involves skin, nails and mucous membrane and is persistent. It has also been suggested that familial cause might be environmental and related to infection, rather than to genetics.
- *Infection*—a bacterial etiology may be there but results are not confirmed. Spirochetes and rod-like bodies resembling bacteria have also been detected.
- *Psychogenic factor*—a relationship of lichen planus with stress is quoted and neurogenic basis is suggested. Observation mostly in nervous and highly stressed persons is associated with emotional upset, over work and some form of mental strain.
- *Habits*—oral lichen planus have shown association with tobacco habit. Chewers of tobacco and betel have increased prevalence of oral lichen planus. Smoking may play a role in initiating oral lichen planus of plaque type.
- *Miscellaneous*—occurrence of lichen planus is also suggested in association with deficiency of vitamin B₁, B₆ and C, electric potential difference, anemia and patients with secondary syphilis. It can also occur in some cases due to trauma and malnutrition. Exacerbation of lichen planus also correspond to periods of emotional upset, overwork, anxiety, hysteria attack, depression and some form of mental strain.

Types

- Reticular
- Papular
- Plaque
- Atrophic
- Classical
- Erythematous
- Ulcerative
- Hypertrophic
- Erosive
- Bullous
- Hypertrophied
- Annular
- Actinic
- Follicular
- Linear

Clinical Features

- *Age and sex*—it occurs in adulthood with age range for males as 35-44 years and for females 45-54 years. It has more predilections for females.
- *Site*—it may involve skin, oral and other mucous membranes. About 50% of the patients with skin lesions have oral lesions whereas 25% of all lichen planus have only oral lesions. Other mucosal surfaces including larynx, glans penis, esophagus, stomach, nasal and vulva may get involved.
- *Symptoms*—the chief complaint is usually of intense pruritus. The itching associated with LP usually provokes rubbing of the lesions, rather than scratching.
- *Signs*—the lesions have a characteristic violet hue. They are flat-topped, shiny, polygonal papules and plaques. The surface is dry with thin, adherent scales (**Fig. 12-25**).



Fig. 12-25: Lichen Planus seen on the hand of the patient.

- *Six P* of lichen planus—six 'p's characterize the lesions LP: they are planar, polygonal, purple, pruritic, papules and plaques.
- *Vaginogingival syndrome*—oral lesions coexisting with genital mucosal lesions are known as *vaginogingival syndrome*.
- *Skin lesion*—skin lesions appear as small, angular, flat topped papules, only a few mm in diameter, which may be discrete or gradually coalesce into larger plaque and is covered by fine glistening scales (**Fig. 12-26**).



Fig. 12-26: Lichen Planus seen on hand of patient. It is showing glistening scale (Courtesy Dr Chole).

- **Color**—papules are sharply demarcated from surrounding skin, which appears red but soon takes reddish purple or violaceous blue color. Later, dirty brown color develops. Center of papule may be slightly umbilicated
- **Wickham's striae**—these are very fine grayish lines which cover the papules.
- **Koebner phenomenon**—fresh lesions may appear on scratch marks or at sites of other non-specific trauma.
- **Graham little syndrome**—alopecia occurs in some patients which may be termed lichen planus planopilaris (Graham-Little syndrome). Nail changes, particularly longitudinal ridging and grooving may be seen.
- **Grinspan syndrome**—lichen planus can be associated with Grinspan syndrome which consists of lichen planus, diabetes mellitus and vascular hypertension.
- **Complication**—it includes post-inflammatory hyperpigmentation, malignancy from oral lesions, nail dystrophy and scarring alopecia.

Oral Lichen Planus

- **Sites**—common sites are buccal mucosa (84%) and to lesser extent tongue, lips, gingiva, floor of mouth (Fig. 12-27) and palate.
- **Symptoms**—patient may report with burning sensation of oral mucosa.
- **Appearance**—oral lesion is characterized by radiating white and gray velvety thread-like papules in a linear, angular or retiform arrangement forming typical lacy, reticular patterns, rings and streaks over the buccal mucosa and to a lesser extent on the lip, tongue and palate.
- **Wickham's striae**—tiny white elevated dots are present at the intersection of white lines, called as Wickham's striae.
- **Superimposed candidial infection**—in some cases superimposed candidial infection may present.



Fig. 12-27: Lichen Planus seen in floor of mouth showing radiating striae.

- **Reticular type**—it is most common form and is mostly bilateral. It consists of slightly elevated fine whitish lines that produce lace-like pattern of fine radiating lines, called as Wickham's striae (Figs 12-28A to C). The lesion may present radiating white thread like papules in a linear, annular or retiform arrangement. A tiny white dot is frequently present at the intersection of white lines.



Fig. 12-28A



Fig. 12-28B



Fig. 12-28C

Figs 12-28A to C: Reticular type of Lichen Planus seen on buccal mucosa.

- *Papular*—whitish elevated lesions of 0.5 mm to 1 mm in size, well seen on keratinized areas of oral mucosa. Papules are spaced apart; still close enough to give pebbled white or gray color (**Fig. 12-29**). Sometimes they coalesce. Most often, papules are seen at the periphery of reticular pattern.



Fig. 12-29: Papular type of Lichen Planus seen on lip.

- *Plaque*—it is seen on dorsum of tongue and buccal mucosa. In case of plaque of tongue, disappearance of the tongue papillae is seen. It spreads in concentric peripheral growth. It consists of either pearly white or grayish white plaque. Such plaques generally range from slightly elevated and smooth to slightly irregular form (**Fig. 12-30**).
- *Atrophic form*—it appears as smooth, red, poorly defined area, often but not always, with peripheral striae evident. The attached gingiva is frequently involved in this form of lichen planus is called desquamative gingivitis pattern (**Fig. 12-31**). At the margins of atrophic zones,



Fig. 12-30: Plaque type of Lichen Planus showing slightly elevated lesion.

whitish keratotic striae are usually evident, radiating peripherally and blending into surrounding mucosa. The gingiva tends to show patchy distribution over all the four quadrants in a relatively symmetrical pattern. It is always symptomatic with complain of pain and burning in the areas of involvement.



Fig. 12-31: Atrophic form of Lichen Planus showing involvement of attached gingiva.

- *Bullous form*—it consists of vesicles and bullae which are short lived. These upon rupturing, leave an ulcerated extremely uncomfortable surface (**Fig. 12-32**). The most common site is buccal mucosa especially into posterior and lateral margins of tongue. It is often associated with striated or keratotic component.
- *Hypertrophic form*—it appears as well circumscribed, elevated white lesion resembling leukoplakia.
- *Annular form*—it appears as round or ovoid, white outline with either pink or reddish pink center.



Fig. 12-32: Bullous form of Lichen Planus showing ulceration in posterior area.

- *Pigmented lichen planus*—in many cases, lichen planus is associated with pigmentation (**Fig. 12-33**).



Fig. 12-33: Lichen Planus associated with pigmentation in on buccal mucosa.

- *Malignant potential*—the incidence of malignant transformation ranges from 0.4% to 12.3%. In India, the incidence of malignant transformation is 0.4%. Carcinoma development is more common in women than in men. Atrophic, erosive and ulcerative lesions showing erythroplakic component and tobacco chewers are indicated to be more cancer prone.

Clinical Variation of Lichen Planus

In many cases, the clinical picture may differ from the classical one, the variations being in morphology and configuration or there may be modifications of clinical features by the site of involvement.

Variations in morphology

- *Hypertrophic lichen planus*—this may occur as a sole manifestation or as a part of a more generalized subacute disease. The lesions are elevated, warty, pigmented plaques typically occurring on the shin or around the ankle. These lesions tend to be persistent.
- *Atrophic lichen planus*—the lesions are few in number. There are depressed, pigmented lesions.
- *Follicular lichen planus*—it is also known as *lichen planopilaris*, this may accompany typical lesions or may be the sole morphologic type. There are small lesions centered on hair follicles. Lesions on scalp may result in alopecia. The combination of follicular LP with scarring alopecia of scalp and non-scarring alopecia of axilla and pubis or other areas is known as *Graham-Little syndrome*.
- *Actinic lichen planus*—described mainly from the Middle East and India, these lesions are common on the face and present as hyperpigmented patches with a surrounding zone of hypopigmentation.
- *Bullous lichen planus*—vesicles and bullae may occur on typical lesions of LP in this variant, due to severe basal cell degeneration induced by the inflammatory process.
- *Ulcerative lichen planus*—chronic ulcers occur on the feet. It may accompany oral ulcer and loss of nails.
- *Lichen planus pigmentosus*—described mainly from India and Middle East, these lesions present with deeply pigmented macules on the face and extremities.

Variations in shape:

- *Linear lichen planus*—this may follow marks of injury by koebnerization, or may occur spontaneously in long linear arrays or rarely in a dermatome in a zoster-like fashion.
- *Annular lichen planus*—these lesions have central depressed areas with raised margins. Typically occurs on penis.
- *Guttate lichen planus*—large number of drop-shaped lesions.

Variation by site

- *Oral*—oral lesions frequently occur in lichen planus.
- *Genital*—annular plaques often occur on glans penis, vulval or vaginal lesions, often presenting with chronic ulcers, may rarely occur.
- *Nails*—nail dystrophies may occur in lichen planus. The various changes that may occur are: thinning of nail plate, longitudinal ridging, onycholysis, subungual hyperkeratosis and pterygium formation (fusion of proximal nail fold with the nail plate).
- *Palms and soles*—lichen planus of these locations lack the typical color; they are yellowish rather than violaceous papules and plaques.

- *Scalp*—follicular lesions with scarring alopecia are seen at this site.

Special variant

- *Drug-induced lichenoid reactions*—a number of drugs may induce development of lesions that clinically and histologically resemble idiopathic lichen planus. The principal offenders are: gold, penicillamine, beta-blockers, anti-malarials, captopril, lithium, carbamazepine, chlorpromazine, thiazides, methyldopa, and NSAIDs.
- *Lichen planus pemphigoides*—bullous lesions occur on lesions as well as normal skin. This appears to be a variant with combined features of lichen planus and bullous pemphigoid.
- *Lichenoid contact dermatitis*—contact with color film developer containing paraphenyldiamine may give rise to lichen planus like lesions on hands. Amalgams in dental filling material may also give rise to lichenoid oral lesions.
- *Lichen planus-lupus erythematosus overlap*—lesions with atrophic center violaceous border occur mainly on hands and feet. Immunohistologic evidence of both lichen planus and lupus erythematosus, are present.

Diagnosis

- *Clinical diagnosis*—the interlacing white striae appearing bilaterally. Presence of Wickham striae and Koebner phenomenon is also diagnostic.
- *Laboratory diagnosis*—there is hyperorthokeratosis, hyperparakeratosis, acanthosis with intercellular edema of spinous cells. Biopsy also shows *civatte bodies* in spinous and basal cell layers and lamina propria. *Saw tooth appearance* of the rete pegs is seen.
- *Immunofluorescent study*—positive for direct immunofluorescence reaction with IgA, IgM, IgG antisera. Most constant feature is presence of sub-epithelial deposits of fibrinogen and antigenically related substance, which can be stain by anti-fibrinogen antisera.

Differential Diagnosis

- *Leukoplakia*—men are more commonly affected as compared to lichen planus. Also in lichen planus, Wickham's striae are present.
- *Candidiasis*—pseudomembrane can be rubbed off in case of candidiasis.
- *Pemphigus*—characteristic clinical white striation of lichen planus are usually evident in cases. Diagnosis of pemphigus can be made by microscopic finding of acantholysis.
- *Lupus erythematosus*—Not in fine reticular pattern, but have much broader dimension. Flaky and feathery appearance of lupus lesion.

- *Drug induced lesion*—history of drug administration.
- *White sponge nevus*—it is seen at birth and puberty; lichen planus is seen over the age of 30 years.
- *Ectopic geographic tongue*—red center with slightly raised margins which rapidly alter.
- *Cheek biting*—history of trauma is usually given.
- *Lichenoid drug reaction*—initiate a course of systemic administration of steroids, if no improvement occurs in 2 weeks, lichenoid reaction will be the working diagnosis.

Management

- *Removal of cause*—the causative factor is removed and this may lead to resolution of lesion subsequently. This can be particularly applicable to lichenoid drug eruption.
- *Steroids*—in most patients with erosive and ulcerative lesion steroids are commonly used. The rationale behind their use is their ability to modulate inflammation and immune response. Topical and intralesional routes are used when systemic steroids are contraindicated. These routes are useful when the patient refuses needle injection or when treating painful gingival sites, where injection delivery is impossible. The topical, injectable and systemic routes are used when there is no systemic contraindication and a full steroid dose is required.
- *Steroid spray*—small and moderately sized painful lesions can be treated with *beclomethasone dipropionate spray*, *triamcinolone acetonide* in gel or cream base.
- *Steroid coating in soft custom tray*—in case of some painful gingival lesions topical steroids may be applied using soft custom trays by coating steroids on the undersurface of the tray. This tray anchors to the dental arch while covering the painful gingival lesion.
- *Topical delivery regimen*—this earlier regime called topical delivery QID, for 3 or more weeks, once a week intralesional injection, for 3 weeks (usually -0.5 ml – 1.5 ml) and systemic steroid (prednisolone 5 mg tab) in tapering doses of 30 mg/day for the first of 3 weeks, 15 mg/day for second week and 5 mg/day for third and final week. Intralesional injections which are used are *methylprednisolone* 40 mg/ml and *triamcinolone acetonide* 10 mg/ml.
- *Topical application*—topical application of *fluocinolone acetonide* for 4 weeks is also effective in curing the disease (**Fig. 12-34**).
- *Combination of prednisolone and levamisole*—another regime consisting of prednisolone and levamisole has also been tried successfully recently. This systemic regime calls for prednisolone 5 mg and levamisole 50 mg tablets for first three days of rest and this schedule to be followed for two to three more weeks.



Fig. 12-34: Post-treatment photograph of Lichen Planus (Courtesy Dr Tapasya Karamore).

- **Topical application of antifungal agent**—it is given when candidiasis superimposed lichen planus. Steroid therapy is routinely accompanied by antifungal treatment as steroid therapy tends to generate an oral fungal infection. The prophylactic antifungal therapy usually consists of clotrimazole oral torches. Nystatin and ketoconazole can also be used.
- **Vitamin A (retinoid) analogue**—retinoids are useful usually in conjunction with topical corticosteroids as adjunctive therapy either topically or systemically. This is because of their anti-keratinizing and immunomodulating effects. They are particularly effective against keratinized reticular and plaque variants.
 - **Topical vitamin A acid cream (0.1%)** Tretinoin, beta altransretinoic acid, systemic etretinate and systemic and topical isotretinoin are also useful in resolution of the lesions, but withdrawal of the medication leads to rapid recurrence to the lesion very oftenly.
 - **Side effect**—the side effects of retinoids are more and it includes foci of erythema during and after the topical treatment. For systemic retinoids, it includes liver dysfunction, cheilitis and dryness of mucous membrane.
 - **Temarotene**—a new systemic retinoid; temarotene, has been reported effective and free of side effects, other than a slight increase in liver enzymes.
- **Cyclosporine**—it is a selective inhibitor of CD₄ helper T lymphocytes that is used systemically to achieve immunosuppression. It can be used both, topically and systemically. The lesion shows complete healing with no recurrence following 8 weeks of systemic cyclosporine 8 mg/kg/day. Oral lesions can be treated using cyclosporine as a rinse and expectorant. It is used as 5 ml rinse, TID, for 8 weeks.

- **Surgical therapy**—it is indicated when conventional methods fail in ulcerative lesion and small solitary lesions. In some cases, cryosurgery and cauterization have also been tried.
- **Psychotherapy**—emotional status of the patient is important in the development of this disease. In some cases, the lesion may regress when the patient is made aware of psychogenic implication of the condition and the nature of emotional stress is understood. When the lesions are asymptomatic and there is no source of emotional distress, it is often advisable to refrain from therapy as failure to eradicate the lesion by medication may trigger the patient into becoming fearful of cancer. Tranquilizers have also been tried to reduce anxiety.
- **Dapsone therapy**—dapsone diamino-diphenylsulphone is an antibacterial sulphone. It is postulated that this particular agent may help to control the lymphocyte mediated progress of lichen planus by modulating the release of inflammatory or chemotactic factor for mast cells or neutrophils. It is used in severe form of erosive lesions.
- **PUVA therapy**—In this form of therapy, psoralens and high intensity long wave ultraviolet A (PUVA) light is used as a therapeutic agent. The lesion shows improvement during and immediately after the treatment.
- **Fluocinonide**—fluocinonide in an adhesive base can be useful in treating lichen planus.
- **Symptomatic**—it can be provided by topical analgesic, topical anesthetic and anti-histaminic rinse.
- **Others**—apart from above mentioned therapies various other agents are also used in treatment of lichen planus. It includes vitamin B complex, heavy metal preparation such as those containing bismuth, arsenic and mercury, antibiotics such as penicillin, doxycycline, radiation, chloroquine therapy and immunization therapy.

Table 12-1: Treatment options

Group	Treatment
Group I—Lichen Planus of reticular or atrophic variety without symptoms	• No treatment is required • Regular follow-up • Diazepam for anxiety
Group II—Lichen Planus of reticular or atrophic variety except with mild to moderate pain and burning	• Local application of benzocaine 10% for burning • 2 mg of diazepam • Topical corticosteroid • Intralesional steroid for quicker relief • Regular follow-up
Group III—erosive Lichen Planus with or without symptoms	• Immediate biopsy • Local control of pain • Intralesional steroid • If infected, antibiotic
Group IV—lichenoid reaction	• Discontinue the offending drug • Local application of benzocaine

Erosive Lichen Planus

It presents as chronic multiple oral mucosal ulcers, which occur when there is extensive degeneration of basal cell layer of epithelium.

Etiology

- *Drug therapy* like NSAIDs, hydrochlorothiazide, penicillamine, angiotensin converting enzyme inhibitors.
- *Chronic hepatitis*—underlying medical disorders like chronic hepatitis.
- *Dental restoration*—reaction to dental restorations.
- *Graft versus host disease*—graft versus host disease due to bone marrow transplantation.
- *Stress*—emotional stress can lead to erosive lichen planus.

Clinical Features

- *Age and sex*—female to male ratio is 2:1. Average age is 50 years. It is primarily a disease of whites, but may be seen in blacks.
- *Sites*—common site is buccal mucosa and lingual mucosa.
- *Symptoms*—patient complains of burning sensation and pain.
- *Appearance*—after rupture of vesicles, eroded or frankly ulcerated lesion are seen which appears as a raw painful areas (Fig. 12-35). Lacy white pattern may be present.
- *Size and shape*—eroded and frankly ulcerated lesions are irregular in size and shape and appear as raw and painful areas (Fig. 12-36).
- *Surface*—the surface is generally granular and brightly erythematous and may bleed upon slight provocation or manipulation.
- *Signs*—a fibrinous plaque or pseudomembrane may be seen over erosion, while later is significant.
- *Malignant potential*—malignant potential is 1 to 25%.



Fig. 12-35: Ulcerated area seen on buccal mucosa in erosive Lichen Planus.

- *Healing*—present for a week to month and heal in periods of 10 days to 2 weeks.



Fig. 12-36: Irregular shaped ulcerated lesion with lacy white pattern at the periphery is seen in erosive Lichen Planus.

Diagnosis

- *Clinical diagnosis*—ulcerative lesion with white lacy pattern is diagnostic indicator of erosive lichen planus.
- *Laboratory diagnosis*—In the erosive form, the epithelium is completely missing or only remnants of epithelial tissues are seen. It shows classical feature of lichen planus, i.e. hydropic degeneration of basal cell layer with juxtaepithelial inflammatory cell infiltrate.

Differential Diagnosis

- *Speckled leukoplakia*—surface is speckled, as compared to erosive lichen planus where it is granular.
- *Squamous cell carcinoma*—borders of the lesion are ill defined and surface is irregular.
- *Lichenoid drug reaction*—history of drug administration.
- *Electrogalvanic mucosal lesion*—dissimilar restorations present in the oral cavity.
- *Psoriasis*—Auspitz's sign is positive.
- *Atrophic candidiasis with keratotic borders*—can be confused with an erosive lichen planus especially when the candidial focus is circumscribed by a keratotic border; proof of cause is searched for typical lichen planus.
- *Cicatricial pemphigoid*—eye lesions are present in cases of cicatricial pemphigoid.
- *Erythema multiforme*—onset less acute, whitish lichen design on the borders of erosion and skin changes occur in case of lichen planus.
- *Desquamative gingivitis*—the gingival surface is smooth and shiny and there is patchy distribution of red and gray areas.
- *Bullous skin lesion*—lichen planus lesion yields a negative blister induction test.

- *Recurrent aphthae*—whitish changes in border lesion present in case of lichen planus, which is absent in the case of recurrent aphthae.
- *Primary syphilitic lesion*—painless indurated edema, painless swollen lymph nodes.

Management

It is same which is given for other type of lichen planus.

Graft-versus-Host Disease (GVHD)

It occurs in a person who receives allogenic bone marrow transplantation. This occurs when in some cases in spite of HLA matching donor person rejects allogenic bone marrow transplantation and patient can suffer from variety of signs and symptoms.

Clinical Features

- *Acute GVHD*—it is seen after one or two weeks of allogenic bone marrow transplantation. The skin lesion ranges from mild rash to diffuse severe sloughing. It is also associated with diarrhea, nausea, vomiting, abdominal pain, liver dysfunction.
- *Chronic GVHD*—it may develop after 3 months of duration. Skin lesion resembles as lichen planus. Some patients may notice features similar to lupus erythematosus, Sjögren syndrome.

Oral Manifestation

- *Site*—tongue, labial mucosa, and buccal mucosa are most commonly involved site.
- *Symptoms*—burning sensation and mucosal discomfort is present. Patient also complains of xerostomia.
- *Lichen planus like lesion*—oral lesion of graft versus host resistance resembles lichen planus (Fig. 12-37).



Fig. 12-37: Lichen Planus type lesion seen in patient with allogenic bone marrow transplantation.

- *Sign*—in some cases patient may notice mucocele showing signs of salivary gland involvement.

Diagnosis

- *Clinical diagnosis*—lichen planus type of lesion with history of allogenic bone marrow transplantation will diagnose this condition.
- *Laboratory diagnosis*—there is abnormal deposition of collagen. Other features are similar to lichen planus.

Management

- *Topical steroids*—topical steroids will facilitate healing of oral ulceration.
- *Topical anesthetics gel*—it is also used to control burning sensation of oral cavity.
- *Thalidomide*—this is useful drug in cases of chronic GVHD which are resistant to standard therapy.
- *Prevention*—prophylactic therapy with immunomodulator and immunosuppressive drugs such as cyclosporine and prednisone should be given to prevent GVHD. Doses the above drugs should be increased if GVHD occurs.

Lichenoid Reaction

The unifying feature of lichenoid reaction and lichen planus is the similar clinical and light microscopic appearance of the skin and mucous membrane lesions. Lichenoid reactions were differentiated from lichen planus on the basis of association of lichenoid reaction with administration of a drug or a systemic disease and their resolution, when the drug was discontinued.

Etiopathogenesis

- *Disorders*—lichen planus, lupus erythematosus, erythema multiforme, fixed drug eruptions, secondary syphilis and graft-versus-host reaction.
- *Drugs and chemical*—the tissues of a person with diathesis react in a special way to certain extrinsic stimuli, making it more susceptible to certain diseases. Drugs act to increase temporarily the specific antigenic stimulus and hence increase the reaction. If the drug is withdrawn at a later time, the antigenic stimulus is reduced, followed by reduction in clinical severity. It is suggested that drugs that are known to induce lichenoid response, act as agents who amplify the disorder, rather than induce it. To implicate a drug responsible for a lichenoid reaction can be difficult as there is no specific test for it. Association between dental filling material and lichen planus has also been suggested. Following drugs can cause lichenoid reaction.

- *Antimicrobial*—dapsons, para-aminosalicylate, streptomycin and tetracycline.
- *Anti-parasitic*—antimony compounds, organic arsenical, chloroquine and quinacrine.
- *Anti-hypertensive*—chlorothiazide, hydrochlorothiazide, labetalol, mercurial diuretic, methyl dopa and practolol. Angiotensin converting enzyme inhibitors can also cause lichenoid reaction.
- *Anti-arthritis*—aurothioglucose, colloidal gold, gold sodium thiomalate and gold sodium thiosulfate.
- *Oral hypoglycemic agents of sulfonylurea type*—chlorpropamide and tolbutamide.
- *Miscellaneous drugs*—iodides, penicillamine, quinine sulfate and copper in dental casting alloy.

Clinical Features

- *Lichenoid mucositis*—this is a term used for lesion of mucosa similar to lichen planus due to drugs.
- *Lichenoid dermatitis*—when lesion presents on the skin it is called as lichenoid dermatitis (**Figs 12-38A and B**).
- *Lichenoid foreign body gingivitis*—this occurs due to foreign material embedded in gingiva causing host response.



Figs 12-38A and B: Lichenoid dermatitis seen on hand of the patient (Courtesy Dr Pincha).

Diagnosis

- *Clinical diagnosis*—lichen planus type lesion with drug history will diagnose lichenoid reaction
- *Laboratory diagnosis*—lichenoid drug reaction may show deep as well as superficial dermal lymphocyte infiltrates rather than the classical band-like infiltrate of the lichen planus.

Management

Majority of it are resolved after discontinuation of drug.

Oral Submucous Fibrosis (OSMF)

It is a chronic and high risk precancerous condition. The condition was prevalent in the days of Sushruta (600 B.C.), a great practitioner of ancient medicine where he labeled this condition as 'Vidhari'. After lapse of many years, Schwartz (1952) was the first person to bring this condition again into limelight. He described the condition as '*atrophica idiopathica mucosae oris*'. After that, the condition has also been described as idiopathic scleroderma of mouth, idiopathic palatal fibrosis and sclerosing stomatitis.

Definition

- An insidious, chronic disease affecting any part of the oral cavity and sometimes pharynx. Although occasionally preceded by and/or associated with vesicle formation, it is always associated with juxtaepithelial inflammatory reaction followed by fibroelastic changes of lamina propria, with epithelial atrophy leading to stiffness of oral mucosa and causing trismus and inability to eat.

Epidemiology

The disease is very common in India, Indian subcontinent and other Asian people. The prevalence rate of oral submucous fibrosis in India, Burma and South Africa ranges from 0 to 1.2%. In India, overall incidence is about 0.2% to 0.5%. Its incidence is high in southern parts of India, where the incidence of oral cancer is also high.

Etiopathogenesis

- *Chillies*—the use of chillies (*capsicum annum* and *capsicum frutescence*) have been thought to play an etiological role in oral submucous fibrosis. Capsaicin is the active ingredient of chillies. It is the vanillylamide of 8-methyl-6-nonenic acid, which is the active irritant of the chillies.
- *Tobacco*—it is a known irritant and causative factor in oral malignancy. It may act as a local irritant in oral submucous fibrosis.

- *Lime*—lime is used with betel nut for chewing. It causes local irritation and damage to the mucosa with vesicle and ulcer formation in susceptible individuals. It acts as a local irritant.
- *Betel nut*—the term areca nut is used to denote the unhusked whole fruit of the areca nut tree and term betel nut is used exclusively to refer to the inner karnel or seed which is obtained after removing husk. The betel nut has psychotropic and antihelmintic property due to presence of areca alkaloids, predominantly arecoline. These alkaloids have powerful parasympathetic properties which produce euphoria and counteract fatigue. Areca nut is found to contain different types of alkaloids like arecoline, arecadine, arecalidine, guvacoine, guvacine and isoguvacine. The pathogenesis of oral submucous fibrosis caused by betel nut is as follows:
 - *Tannic acid and arecoline content*—in a habitual betel nut chewer, oral submucous fibrosis may be caused by the amount of tannic acid contained in the betel nut, the influence of mixed calcium powder and the conditional action of arecoline content in betel nut, affecting the vascular supply of oral mucosa and causing neurotropic disorder.
 - *Formation of areca nut specific nitrosamine*—nitrosation of arecoline leads to the formation of areca nut specific nitrosamine namely nitrosoguvacoline, nitrosoguvacine and 3-methyl nitrosomino piperidine, which alkylate DNA
 - *Metabolism of areca nut specific nitrosamine*—metabolism of these areca nut specific nitrosamine will lead to formation of cyanoethyl, which adducts with o'-methyl guanine in DNA. Prolonged exposure to this irritant leads to malignant transformation.
 - *Dual action of areca nut*—recently suggested that pathogenesis of oral submucous fibrosis is by dual action of areca nut. It is suggested that arecoline stimulates fibroblastic proliferation and collagen synthesis. The flavanoid catechin and tannins are also components of areca nut and they stabilize the collagen fibrils rendering them resistance to degradation by collagenase. The attendant trismus is the result of juxtaepithelial hyalinization and secondary muscle involvement. Glycogen consumption is physiologically related to cellular activity. Over activity of muscle results in excessive glycogen consumption, leading to glycogen depletion. The increased muscle activity and diminished blood supply, following connective tissue changes; owing to extensive oral submucous fibrosis leads to muscle degeneration and fibrosis.
 - *Nutritional deficiency*—the disease is characterized by repeated vesiculations and ulcerations of oral cavity. A sub-clinical vitamin B complex deficiency has been suspected in such cases. The deficiency could be precipitated by the effect of defective nutrition due to impaired food intake in advanced cases and may be the effect, rather than the cause of the disease.
- *Defective iron metabolism*—microcytic hypochromic anemia with high serum iron have been reported in submucous fibrosis but as such, there is no definite proof available to support this cause effect relation.
- *Bacterial infection*—streptococcal toxicity is also a factor in etiology of oral submucous fibrosis, as in some other collagen disorder such as rheumatic disease. *Klebsiella rhinoscleromatis* may be a factor in cause of submucous fibrosis.
- *Collagen disorders*—oral submucous fibrosis is thought to be localized collagen disease of oral cavity. It is linked to scleroderma, rheumatoid arthritis, Dupuytren's contracture and intestinal fibrosis. A link between scleroderma and oral submucous fibrosis has also been suspected on the basis of similarity of histological characteristic.
- *Immunological disorders*—raised ESR and globulin levels are indicative of immunodeficiency disorder. Serum immunoglobulin levels of IgA, IgG and IgM are raised significantly in oral submucous fibrosis. These raised levels suggest an antigenic stimulus in the absence of any infection. Circulating auto-antibodies are also present in some cases of oral submucous fibrosis.
- *Altered salivary composition*—the study of saliva in cases of oral submucous fibrosis have shown increased pH, increase in salivary amylase, low levels of calcium, increase in alkaline phosphatase and potassium and normal levels of salivary immunoglobulin. The fibrin precipitating factor in saliva has been attributed to the increased plasma fibrinogen. This is likely due to increased dietary content of fibrin.
- *Genetic susceptibility*—the familial occurrence of oral submucous fibrosis has also been reported (**Fig. 12-39**).

Clinical Features

- *Age and sex distribution*—it affects both sexes. The age group varies, although majority of patients are between 20 and 40 years of age.
- *Site distribution*—the most frequent location of oral submucous fibrosis is the buccal mucosa and the retro-molar areas. It also commonly involves soft palate, palatal fauces, uvula, tongue and labial mucosa. Sometimes, it involves the floor of mouth and gingiva.
- *Prodromal symptoms*—the onset of the condition is insidious and is often of 2 to 5 years of duration. The most common initial symptom is burning sensation of oral mucosa, aggravated by spicy food, followed by either hypersalivation or dryness of mouth. Vesiculation,



Fig. 12-39: Familial oral submucous fibrosis (OSMF) affecting daughter of patient who is also having OSMF.

ulceration, pigmentation, recurrent stomatitis and defective gustatory sensation have also been indicated as early symptoms.

• *Late symptoms*

- *Trismus*—gradual stiffening of the oral mucosa occurs in few years after the initial symptoms appear. This leads to inability to open the mouth completely (Fig. 12-40).
- *Difficulty in tongue protrusion*—later on, patients experience difficulty in protruding the tongue.
- *Difficulty in swallowing*—when the fibrosis extends to pharynx and esophagus, the patient may experience difficulty in swallowing the food.
- *Referred pain*—referred pain in the ears and deafness, due to occlusion of Eustachian tube and a typical nasal voice has been reported.



Fig. 12-40: Reduced mouth opening in oral submucous fibrosis (OSMF) (Courtesy Dr Suwas Darvekar).

- *Blanching of mucosa*—the most common and earliest sign is blanching of mucosa, caused by impairment of local vascularity (Fig. 12-41). The blanched mucosa becomes slightly opaque and white. The whitening often takes place in spots so that the mucosa acquires a marble like appearance. Blanching may be localized or diffuse, involving greater part of the oral mucosa or reticular, in which blanching consists of blanched area with intervening clinically normal mucosa, giving it a lace-like appearance.



Fig. 12-41: Blanched palate seen in OSMF.

- *Betel chewer mucosa*—it is brownish red discoloration of mucosa with irregular surface that tend to desquamate.
- *Fibrous band*—As disease progresses the mucosa becomes stiff and vertical fibrous band appears. This band can be palpate easily and feel rough on palpation.
- *Lips features*—mucosa is blanched, becomes rubbery and is characterized by the presence of circular bands around the rima oris like a thin band (Fig. 12-42). In severe labial



Fig. 12-42: Circular band seen around the rima oris in patient with oral submucous fibrosis (OSMF).

involvement, the opening of mouth is altered to an elliptical shape (elliptical rima oris), lips become leathery and it become difficult to evert them.

- *Buccal mucosa*—the affected mucosa becomes coarse, blanched and inelastic. In advanced cases, the mucosa becomes tough and leathery with numerous vertical fibrous bands (Fig. 12-43).



Fig. 12-43: Blanched fibrous band seen in buccal mucosa (Courtesy Dr Bhaskar Patle),

- *Soft palate (49%) and uvula*—involvement of soft palate is marked by fibrotic changes and a clear delineation of the soft palate from hard palate. The mobility of soft palate is restricted. Uvula, when involved, is shrunk and in extreme cases it becomes bud-like or hockey stick appearance (Figs 12-44 and 12-45).



Fig. 12-44: Bud-like appearance seen of uvula in oral submucous fibrosis (OSMF) (Courtesy Dr Bhaskar Patle).

- *Palatal fauces*—In the soft palate the bands radiate from pterygomandibular raphe to the anterior faucial pillars. The faucial pillars become thick and short and tonsils may get pressed in between fibrosed pillars.



Fig. 12-45: Hockey stick appearance of uvula seen in oral submucous fibrosis (OSMF) (Courtesy Dr Parate).

- *Tongue*—The initial change is depapillation, usually in the lateral margins. Tongue becomes smooth (Fig. 12-46); its mobility, especially in protrusion, becomes impaired. Patient cannot protrude the tongue beyond the incisal edges.



Fig. 12-46: Smooth tongue appears due to depapillation in case of oral submucous fibrosis.

- *Floor of mouth*—when floor of mouth is affected, it becomes inelastic.
- *Gingiva*—when affected, it becomes fibrotic, blanched and inelastic.
- *Associated features*
 - *Pigmentation*—hyperpigmentation or occasional loss of pigmentation is very common in association with oral submucous fibrosis. Many times pigmentation changes in vermillion border are so striking that this disease can be suspected even before examining the patient.
 - *Vesicle*—it is usually found in areas of redness in the soft palate, the anterior faucial pillar, buccal mucosa

or the mucosal surface of lip, particularly the lower lip. The vesicles are painful and they soon rupture leaving behind superficial ulceration. Often there is history of vesiculation following the intake of spicy food, suggesting an allergic reaction to spicy food.

- *Ulceration*—ulceration often develops in the course of disease, particularly in advanced cases. In advanced cases, epithelium becomes atrophic, fragile and vulnerable to ulceration (**Fig. 12-47**).



Fig. 12-47: Ulceration seen in oral submucous fibrosis.

- *Petechiae*—these are small raised reddish blue spots which sometimes occur in oral submucous fibrosis. It may be few or many. They occur most commonly on tongue and the labial and buccal mucosa. The petechiae are transient in nature and do not require any specific treatment.

Clinical Stages of Oral Submucous Fibrosis (Table 12-2)

- *Stage of stomatitis and vesiculation*—this is the earliest stage and is characterized by recurrent stomatitis and vesiculation. Patient complains of burning sensation in the mouth and inability to eat spicy food. The examination reveals vesicle formation particularly on the palate. They may rupture and superficial ulceration may be seen, which may cause painful mastication. Some amount of fibrosis is seen in this stage and mucosa shows whitish streaks. An occasional granulating red spot may be seen on the palate.
- *Stage of fibrosis*—the patient complains of stiffness and inability to open the mouth completely. As the disease progresses, there is difficulty in blowing out the cheeks. Tongue movement becomes restricted and protrusion of tongue is difficult. Complaints of pain in the ear may

Table 12-2: Clinically grading of oral submucous fibrosis according to its severity

Features	Grade I incipient (Very early stage)	Grade II (Mild)	Grade III (Moderate)	Grade IV (a) (Advanced stage)	Grade V (b) (Advanced pre-malignant and malignant change)
Symptoms	Burning sensation, dryness of mouth, vesicle or ulceration	Burning sensation, dryness of mouth	Burning sensation, dryness of mouth	Burning sensation, dryness of mouth	All classical sign of oral submucous fibrosis and it is associated with leukoplakia and Lichen Planus
Spicy food	Irritation with spicy food	Irritation with spicy food	Irritation with spicy food	Irritation with spicy food	
Mucosal color	No changes in mucosal color	Mucosa is blanched and loses its elasticity	Blanched opaque leather-like mucosa	Blanched opaque leather-like mucosa	
Fibrosis	No fibrosis, bands palpable	No clear-cut fibrotic bands	Vertical fibrotic bands on buccal mucosa making it stiff	Thick fibrosed bands occurring on both the buccal mucosa in retromolar area and at pterygomandibular raphe	
Mouth opening	Mouth opening normal (44 mm)	Slight restriction of mouth opening (26-35 mm)	Considerable restriction of mouth opening (15-25 mm)	Very little mouth opening (2-15 mm)	
Tongue	Tongue protrusion normal	Tongue protrusion normal	Tongue protrusion not much affected	Restricted tongue protrusion	
Eating and speaking	—	—	Difficulty in eating and speaking	Eating and speech very much impaired	
Oral hygiene	—	—	Poor oral hygiene	Very poor oral hygiene	

occur occasionally. Speech may become muffled and indistinct due to restriction of jaw movement. Due to restricted palatal movement, a nasal twang may occur in speech. The examination reveals increased fibrosis of submucosal tissue, which appears blanching and white. The lip and cheek become stiff and the vestibule of mouth is gradually reduced and almost obliterated. This causes difficulty even in introducing the examining fingers in between lips, cheeks and teeth in advanced cases. The palate shows blanching and fibrosis which cause shortening and disappearance of uvula in advanced cases. The fibrotic bands extending from palate to tongue cause strangulation of tonsils and they appear buried in the faucial pillars. The dorsum of tongue shows atrophy of papillae. The mucosa of the floor of mouth beneath the tongue, also show blanching and stiffness.

- *Stage of sequelae and complications*—the patient presents with complaints as described above in stage II. On examination, evidence of whitish leukoplakic changes and rarely an ulcerating malignant lesion may be seen.

Diagnosis

- *Clinical diagnosis*—clinically reduced mouth opening with palpable fibrous band is enough to make diagnosis.
- *Laboratory diagnosis*—oral epithelium is markedly atrophic which exhibits intercellular edema, signet cells, and epithelial atypia. The inflammatory cells are mostly mononuclear; eosinophils and occasional plasma cell may be seen.

Malignant Potential

Atrophic epithelium first becomes hyperkeratotic and later, intracellular edema and basal cell hyperplasia develop eventually, following epithelial atypia with moderate epithelial hyperplasia and then, carcinoma can develop at any time (**Fig. 12-48**).

The WHO Collaborating Centre for Oral Precancerous Lesions has concluded that although oral submucous fibrosis predisposes to cancer, it is not absolutely conclusive. It is highly probable that such relationship does exist. Following facts support this hypothesis:

- The frequency of oral leukoplakia in oral submucous fibrosis patient is 6-8 times higher than in control group.
- Carcinoma patients exhibiting oral submucous fibrosis have a frequency which exceed the 1.2% of submucous fibrosis in general population.
- Immunological alterations observed in oral submucous fibrosis are almost similar to that observed in oral cancer.



Fig. 12-48: Oral submucous fibrosis (OSMF) turning malignant which shows ulcerative growth in the oral cavity (Courtesy Dr Chole).

Management

Restriction of habit/behavioral therapy

The preventive measure should be in the form of stoppage of habit, which can be encouraged through public education. Affected patients should be explained about the disease and its possible malignant potential. Improvement in clinical features like gradual increase in inter-incisal opening has been observed in most of the patients who discontinue the habit.

Medicinal therapy

Supportive treatment

- *Vitamin rich diet*—a vitamin rich diet along with iron preparation is helpful to some extent but has little therapeutic value in relieving trismus.
- *Iodine B complex preparation*—iodine-B-complex preparation (Injection Ranodine) is a combination of iodine preparation with synthetic vitamin B complex. The combination of iodine compound with vitamin B complex is responsible for the stimulation of metabolic process and enzymatic process within the body (oxygen reduction, transamination). Intramuscular injection starts with small doses and continuing with larger doses (2 ml ampule daily). The course of 5 injections is repeated after 7 days. Each 2 ml consists of:
 - *Methyltrioxyethyl iodamine*—progressive increasing doses equivalent to 0, 25, 50, 75 and 125 mg of active iodine
 - *Vitamin B₁*—1.0 mg
 - *Vitamin B₆*—0.3 mg
 - *Vitamin B₂*—0.6 mg
 - *Nicotinamide*—15.0 mg
 - *Calcium pantothenate*—1.0 mg

- *Injection of arsenotyphoid and iodine*—arsenotyphoid is a fibrin dissolving agent.

Steroids

- *Local*—hydrocortisone injection along with procaine hydrochloride injection locally in the area of fibrosis. Injections are given fortnightly. The early cases show good improvement with this therapy.
- *Systemic*—A therapy with hydrocortisone 25 mg tablet, in doses of 100 mg/day is useful in relieving burning sensation without untoward effects. Triamcinolone or 90 mg of dexamethasone can be given. This is supplemented with local injection of hydrocortisone 25 mg at biweekly intervals at the affected site. Increased vascularity of the site is observed, which is attributed to fibrinolytic, anti-allergic and anti-inflammatory action of corticosteroid. The fibrosis is prevented by decreasing fibroblastic production and deposition of collagen.

Placental extract

- *Mechanism*—placentrax is an essential biogenic stimulator. It is suggested that it stimulates pituitary adrenal cortex and regulates metabolism of tissue. It also increases the vascularity of tissues. Its use is based on tissue therapy, which states that “animal and vegetable tissue when severed from the parent body and exposed to unfavorable conditions but not mortal to their existence, undergo biological re-adjustment leading to development of substances in state of their survival to ensure their vitality”. Such tissue extracts when implanted into body stimulate metabolic or regenerative process favoring recovery.
- *Placental extract contains*
 - *Nucleotides*—ribonucleic acid (RNA) and adenosine triphosphate (ATP).
 - *Enzymes*—alkaline and acid phosphatase, glutamic oxaloacetic acid transaminase, glutamic acid and pyruvic acid.
 - *Vitamins*—vitamin E, B, B₆, B₁₂, pantothenic acid, nicotinic acid, biotin PABA and folic acid.
 - *Steroids*—17, ketosteroid.
 - *Fatty acids*—linoleic acid, linolenic acid, palmitic acid.
 - *Trace elements*—copper, selenium, magnesium.
- *Form*—placental extract can be separated into four different factors i.e. aqueous extract, lipid extract, immune gamma globulin and tissue coagulated.
- *Placenta as biogenic stimulator*—only the aqueous extract of placenta acts as biogenic stimulator. It accelerates cellular metabolism, aids in absorption of exudate, and stimulates regenerative process, increases physiological function of organs, produces significant enhancement of wound healing and it has notable anti-inflammatory effect.

- *Dose*—the region affected with submucous fibrosis is divided into five regions. Each region is locally injected around fibrous bands, intra-muscularly, at the interval of 3 days for 15 days. Each time 2 ml solution is deposited. This course can be repeated after a month, if required.

Hyaluronidase

- *Mechanism*—improvement in health of mucous membrane, burning sensation and trismus was observed by using hyaluronidase injection. Hyaluronidase, by breaking down hyaluronic acid (ground substance of connective tissue), lowers the viscosity of intracellular cement substance i.e. hyaluronidase decreases cell formation by virtue of its action on hyaluronic acid, which plays an important role in collagen formation.

Lycopene

- *Content*—it is an antioxidant from tomato extract, along with other previously used antioxidants in the treatment of OSMF.
- *Dose*—tab Lycopene 2000 mcg. The drug will be given for a period of three months duration during which time patient will be reexamined every 15 days.

Vitamin E

- *Mechanism*—the use of vitamin E along with dexamethasone and hyaluronidase injections is thought to produce better results. Vitamin E presumably works by—
 - Preventing the oxidation of essential cellular constituents such as the formation of oxidation product.
 - Protecting against various drugs, metals and chemicals and acts as scavenger of free radical.
 - It may improve the survival of erythrocytes.

Other therapies

- *Vasodilator injection*—vasodilator injection, which relieves the ischemic effect and helps the nutritional and therapeutic measures to reach the affected tissue, with use of fluorouracil an anti-metabolic agent.
- *Injection of interferon gamma*—This is recently discovered therapy. Intralesional injection of interferon gamma improved mouth opening and reduce mucosal burning.

Surgical treatment

Conventional

- *Indication*—surgical treatment is the method of choice; when there is marked limitation of opening of the mouth, in cases where biopsy reveals neoplastic changes and when there is marked trismus and dysphagia. Various authors suggest different approaches.
- *Flap use as graft*—excision of fibrous bands followed by use of tongue flap as a graft gives good results because

tongue flap is highly vascular and resists further fibrosis. The most significant fact is that the tongue mucosa is immune to fibrosis whatever the cause. The aims are to relieve trismus, to prevent further fibrosis and to provide neovascularization of fibrous tissue. It is done in patients who have got mouth opening less than 1.5 cm, in cases of failure of conservative treatment and absence of any other pathology like ulceration, infection and leukoplakia.

- *Implantation of fresh human placenta*—in some cases, submucosal implantation of bits of fresh human placenta into the defect is done following the surgical excision of fibrous bands. The rationale for using placental graft is that they have both, hormonal and mechanical effects. They have stimulant effects because the placenta is homograft that is immunologically competent and rich in steroids, proteins, gonadotrophin, estrogen and progesterone. The graft undergoes local absorption after prolonged period, thus mechanically prevent fibrosis.
- *Nasolabial flap*—the excision of fibrotic bands is followed by reconstruction using bilateral full thickness nasolabial flap.
- *Bilateral palatal flap*—new technique is bilateral palatal flap to cover the exposed area, in combination with bilateral temporalis myotomy and coronoidectomy. After releasing fibrous bands, the anterior border of ramus is exposed. Temporalis tendon and muscle are detached and then coronoidectomy is performed bilaterally.
- *Oral stent*—use of oral stent as adjunct to treatment of oral submucous fibrosis. A stent is an appliance made up of acrylic having posterior vertical stops bilaterally. It is made prior to surgery up to desired increase in mouth opening, to allow the tissue to heal at new height. It is considered especially when the surgical technique is prone to relapse.

Laser

- *Mechanism*—the use of laser for treatment of submucous fibrosis is useful. CO₂ laser surgery offers advantage in alleviating the functional restriction, when compared to traditional surgical technique and subsequent grafting.
- *Technique*—under general anesthesia a CO₂ laser is used to incise the buccal mucosa and vaporize the submucosal connective tissue to the level of the buccinator muscle. Hemostasis is provided by lased surface itself and the mouth opening increases immediately.

Cryosurgery

- *Mechanism*—it is the method of local destruction of tissue by freezing it *in situ*. Open liquid sprays are better suited for mucosal lesion that are extensive and superficial and do not involve the basement membrane.

Oral Physiotherapy

Oral exercises are advised in early and moderately advanced cases. This includes mouth opening and ballooning of mouth. This is thought to put pressure on fibrous bands. Forceful mouth opening have been tried with mouth gag and acrylic surgical screw.

Diathermy

Microwave diathermy is useful in some early or moderately advanced stages. Low current is used (20 watts × 2450 cycles). It acts by physiofibrinolysis of bands. Its value is increased if it is combined with other treatment modalities.

Dyskeratosis Congenita

It is also called as 'Zinssner-Engman-Cole syndrome'. It is a well recognized but rare genokeratosis, which is probably inherited as a recessive characteristic. Mutation in DKC 1 gene will cause disruption in maintenance of telomerase which is critical in determining the normal cellular longevity. Disease manifested has three typical signs: oral leukoplakia, dystrophy of nails and pigmentation of skin.

Clinical Features

- *Age and sex distribution*—it is evident during first 10 years of life. It is almost exclusively seen in males.
- *Nail changes*—nail changes are the first manifestation, becoming dystrophic and shedding some time after the age of 5 years (**Fig. 12-49**).
- *Pigmentation*—grayish brown pigmentations appear in some time which is seen usually on trunk, neck and thigh.
- *Skin*—the skin may become atrophic, telangiectatic and face appears red.



Fig. 12-49: Dystrophy of nails seen in dyskeratosis congenita.

- *Thrombocytopenia*—it is developed during second decade of life.
- *Other features*—other minor manifestations are frail skeleton, mental retardation, small sella turcica, dysphagia, transparent tympanic membrane, deafness, epiphora, eyelid infection, urethral anomalies, small testes and hyperhydrosis of the palms and soles.

Oral Manifestations

- *Sites*—most common sites are tongue and buccal mucosa.
- *Appearance*—it appears as diffusely distributed vesicles and ulcerations, followed by accumulation of white patches of necrotic epithelium and sometimes, superimposed monilial infection. There is also atrophy of tongue (Fig. 12-50).
- *Recurrent ulceration*—after some time, in the age group of 14 to 20 years, there are repeated recurrent ulceration and development of erythroplasia or red mucosal lesion.
- *Erosive leukoplakia*—finally between the age of 20 and 30 years, there is development of erosive leukoplakia and carcinoma.

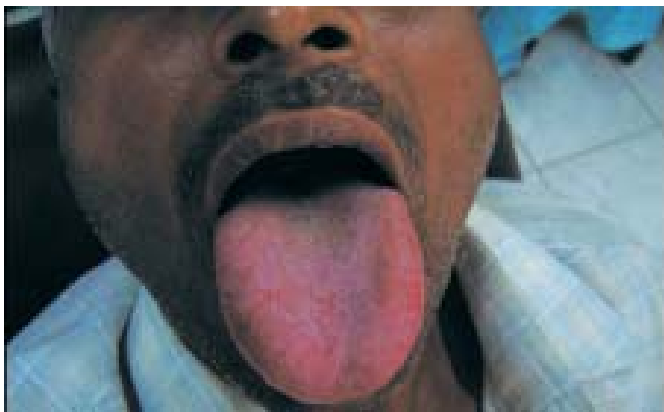


Fig. 12-50: Atrophy and hyperkeratinization seen in dyskeratosis congenita.

Radiographic Feature

- There may be severe periodontal bone loss.

Diagnosis

- *Clinical diagnosis*—leukoplakic type lesion associated with nail changes and pigmentation will diagnose dyskeratosis congenita.
- *Laboratory diagnosis*—the skin lesion shows increased number of melanin containing chromatophores and increased vascularity. There is anemia, leukopenia, thrombocytopenia and pancytopenia.

Management

- *Periodic check up*—periodic check up should be done for evidence of malignant transformation.

Lupus Erythematosus

It is characterized by presence of abnormal antibodies and immune complex.

Types

- *Discoid or cutaneous lupus erythematosus*—if it is confined to skin and mucosa.
- *Systemic lupus erythematosus*—if multiorgan involvement occurs.

Etiology

- *Genetic predisposition*—relative of patients have higher incidences of auto-antibodies, immune deficiency and connective tissue disease. This tendency is greatest among identical twins.
- *Immunological abnormality possibly mediated by viral infection*—immune complex consisting chiefly of nucleic acid and antibody account for majority of the tissue changes.
- *Autoimmune disease*—as these patients develop antibodies to many of their own cells.
- *Endocrine*—there is high incidence in females in pregnancy. This finding suggestive of increased estrogen level.
- *Biochemical increase* in excretion of metabolic products, particularly tyrosine and phenylalanine, in certain SLE patient.

Discoid Lupus Erythematosus

Clinical Features

- *Age and sex*—it occurs in 3rd and 4th decades, female predilection in the ratio of 5:1.
- *Site*—most common sites are face, oral mucosa, chest, back and extremities.
- *Appearance*—it is a circumscribed, slightly elevated, white patch that may be surrounded by red telangiectatic halo.
- *Cutaneous lesion*—cutaneous lesions are slightly elevated, red or purple macules; that are often covered by gray or yellow adherent scales.
- *Carpet track extension*—forceful removal of scale results in 'carpet track extension', which has dipped into enlarged pilosebaceous canals.
- *Peripheral growth*—the lesion increases in size by peripheral growth. Periphery of the lesion appears pink or red, while the center exhibits an atrophic scarred appearance.
- *Butterfly distribution*—butterfly distribution on macular region and across the bridge of the nose.

Oral Manifestations

- *Sites*—most common sites are buccal mucosa, tongue, palate and vermilion border of lip.

- *Symptoms*—there may be burning and tenderness which may be intermittent or disappears if the lesion becomes inactive.
- *Appearance*—it begins as erythematous area, sometimes slightly elevated, but more often depressed, usually with induration and typically with white spots. Occasionally, superficial painful ulceration may occur with crusting or bleeding, but no actual scale formation is seen.
- *Margins*—the margins of the lesion are not sharply demarcated. Fine white striae radiate out from the margins.
- *Central healing*—central healing may result in depression.
- *Lip*—erythematous, atrophic plaque, surrounded by keratotic border may involve the entire lip.

Systemic Lupus Erythematosus

Clinical Features

- *Age and sex*—it occurs in 3rd (female) and 4th (male) decades and has female predilection (8:1).
- *Sites*—it is characterized by repeated remissions and exacerbations with common sites being face, neck, upper arm, shoulders and fingers.
- *Symptoms*—it is manifested by symptoms of fever and pain in the muscle and joints. It may show as itching or burning sensation as well as area of hyper-pigmentation. Severely intensifies after exposure to sunlight.
- *Butterfly distribution of lesion on face*—the cutaneous lesion consists of erythematous patches on the face, which coalesce to form roughly symmetrical pattern over the cheeks and across the bridge of the nose, is called butterfly distribution.
- *Skin lesions*—skin lesions are widespread, bilateral with signs of acute inflammation. This finding helps to differentiate between skin lesions of DLE and SLE.
- *Kidney involvement*—in kidney, fibrinoid thickening of glomerular capillaries producing the characteristic 'wire loop' which may be sufficient to result in renal insufficiency.
- *Heart involvement*—heart may suffer a typical endocarditis involving valves as well as fibrinoid degeneration of epicardium and myocardium.

Oral Manifestation

- *Site*—the most common sites are buccal mucosa, lip and palate.
- *Symptoms*—complain of burning sensation, xerostomia or soreness of mouth.
- *Signs*—lesions similar to DLE, except that hyperemia, edema and extension of lesion is more pronounced.

Greater tendency to bleed and petechiae, suspected ulcerations surrounded by red halo.

- *Appearance*—the intraoral lesion is composed of a central depressed red atrophic area surrounded by 2 to 4 mm elevated keratotic zone that dissolves into small white lines (**Figs 12.51 and 12.52**).
- *Lupus cheilitis*—the lip lesions appear with central atrophic area with small white dots surrounded by keratinized border, which is composed of small radiating white striae. There is occasional ulceration of central area.



Fig. 12-51: Intraoral lesion of systemic lupus erythematosus showing atrophic area (Courtesy Dr Chole).



Fig. 12-52: Same patient (Fig. 12-51) having lesion present on palate (Courtesy Dr Chole).

Diagnosis

- *Clinical diagnosis*—skin lesion with lesion present on oral mucosa which is atrophic and erythematous will suspect lupus erythematosus. Oral and nasopharyngeal ulceration is major diagnostic criteria for SLE.

- *Laboratory diagnosis*—L.E. cell inclusion phenomenon with surrounding pale nuclear mass apparently devoid of lymphocytes. Anemia, leukopenia and thrombocytopenia, with sedimentation rate increased. Serum gamma globulin increased and Coomb's test is positive.
- *Positive lupus band test*—it shows deposition of IgG, IgM or complement component in skin.

Dental Consideration

- *Thrombocytopenia*—it may be sometimes severe. The result of a recent platelet count should be studied before undertaking oral surgery.
- *Bacterial endocarditis*—Libman-Sacks vegetation under the mitral valve may occur in patients with SLE, it can lead to bacterial endocarditis. So patients with SLE should have antibiotic prophylaxis before dental treatment that is likely to cause bacteremia.
- *Exacerbation by drug therapy*—drugs that have been related to exacerbation include penicillin, sulfonamide and NSAIDs with photosensitizing potential.
- *Exacerbation by surgery*—all elective surgeries including dental procedure to be avoided.
- *Susceptibility to shock and infection*—patients with SLE may be taking adrenal suppressive dose of corticosteroids or cytotoxic drugs and hence, they may be susceptible to shock and infection.

Differential Diagnosis

- *Lichen planus*—homogenous picture, no dark erythema and no telangiectasia. Mucosal changes are usually extensive and symmetrical.
- *Lichenoid reaction*—history of drug is always there.
- *Ectopic geographic tongue*—systemic manifestation present is lupus erythematosus, which is absent in ectopic geographic tongue.
- *Psoriasis*—Auspitz's sign is positive.
- *Electrogalvanic lesion*—dissimilar restorations are seen in oral cavity.
- *Leukoplakia and erythroplakia*—lesions tend to maintain same appearance and there are no skin changes.
- *Geographic stomatitis*—no skin changes, mucosal lesions change location rapidly.
- *Benign mucous membrane pemphigoid*—no systemic complain and serology test to be done.

Management

- *Corticosteroid*—systemic lupus erythematosus should be treated by systemic corticosteroids therapy and should be managed by physician. Discoid lupus erythematosus should be treated with topical steroid.
- *Anti-malarial drugs*—anti-malarial drug like hydroxychloroquine combined with nonsteroidal anti-inflammatory agents is also effective treatment modality.

- *Prevention*—patient should avoid exposure to sunlight as this can aggravate the conditions.

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13

Cysts of Jaw

Introduction

Cyst has been known to arise in man (ever since he has teeth) and in certain animals. It takes some odontogenic epithelium plus some unknown initiating factor which stimulates it to proliferate to cause this destructive lesion, which can be found from the mildest form to a greatly disfiguring form.

Cysts of jaws are of great clinical importance, not only because they often attain a large size but also produce facial asymmetry, disturbance of dentition, neurological symptoms and predispose the jaws to fracture but particularly because they have a very high frequency of occurrence.

Definitions

- *By Killey and Key 1966*—this entity constituted an epithelium-lined sac filled with fluid or semifluid material.
- *By some unknown author 1966*—a cyst is an abnormal cavity in hard and soft tissue which contains fluid, semifluid, or gas and is often encapsulated and lined by epithelium.
- *By Kramer in 1974*—pathologic cavity having fluid, semifluid, or gaseous content but not always is lined by epithelium.

General Diagnostic Features of Cysts**Signs**

- *Factors affecting clinical features*—it usually depends on the extent and the dimension of lesion. A small cyst is unlikely to be diagnosed on routine examination because signs will not be demonstrable.
- *Expansion*—as the cyst grows larger, expansion of alveolar bone occurs, usually on the labial and buccal aspect of the jaw in case of a typical odontogenic cyst.

However, expansion of the lingual aspect alone can occur in case of cysts of ramus and third molar area. The expansion of both inner and outer bony wall is often indicative of lesion other than cyst.

- *Enlargement*—as the cyst increases in size, the periosteum is stimulated to form a layer of new bone which is subperiosteal deposition which later forms the outline of the affected portion of the jaw and produce enlargement.
- *Consistency*—at an early stage, the lateral expansion causes smooth, hard, painless prominence but as cyst growth proceeds, the bone covering the center of the convexity becomes thinned and can be indented with pressure. The term elastic is used to describe this particular consistency.
- *Window formation*—in some cases of expanding dental cyst there comes the time when the periosteum appears to be unable to maintain the new bone formation, resulting in an area where there is no bone, usually the summit of the convexity. This is called as ‘window’ formation. Window is more or less round and has a smooth margin which sometimes appears to be very slightly dense at periphery.
- *Egg shell crackling*—later the fragile outer shell of bone becomes fragmented and the sensation imparted and sound produced on palpation over the area is described as ‘egg shell crackling’.
- *Fluctuation*—in advanced cases, these sample pieces of bone disappear and are not replaced by new bone. At this point, the cyst lining lies immediately beneath the mucosa and fluctuation can be elicited.
- *Fluid discharge*—greater distension of the wall of the cyst leads to an eventual discharge of fluid into the mouth. This sequence of events is commonly observed with the progressive enlargement of periodontal and dentigerous cysts.
- *Site*—site of occurrence is also important in considering the diagnostic features. Periodontal cysts may

occur anywhere in the dental arch. Dentigerous cysts however are mainly associated with impacted molars and canines. Fissural cysts are confined to the upper jaw and the solitary bone cyst is virtually assigned to the mandible. Odontogenic keratocysts are most often seen in the lower third molar area extending into ramus.

- *Effect on teeth*—cysts rarely cause loosening of adjacent teeth until they become extremely large. The clinical absence of one or more teeth from normal series without a history of extraction may indicate the presence of developing dentigerous cyst. A single missing tooth may also invite suspicion of existence of odontogenic keratocyst or primordial cyst. The teeth adjoining an odontogenic keratocyst, fissural cysts or solitary cysts are vital. Apical periodontal cyst is associated with non-vital teeth, but in cases of lateral periodontal cysts the involved teeth often are vital.
- *Other features*—a large anterior cyst may extend under the nasal floor and cause distortion of nostril. Involvement of the antrum by an infected cyst may produce signs of maxillary sinusitis. In the jaw a cyst enlarging between teeth can cause their root to diverge and the crown to converge. Afterwards the teeth may tilt over one side.

Symptoms

- *Pain and swelling*—often the first symptom that patient experience is pain and swelling, if the cyst becomes infected. Sometimes the patient notices lump in the sulcus.
- *Anesthesia or paresthesia*—although large mandibular cysts invariably involve the neurovascular bundle and deflect structure into an abnormal position, still it is unusual to find anesthesia of the mental distribution. Since an uninfected cyst grows slowly and exerts a very slight pressure on the neurovascular bundle. However, in cases of acute infection there is a sudden increase in pressure due to pus accumulation in the sac and this can cause neuropraxia of the nerve and the immediate onset of labial paresthesia or anesthesia. After surgical drainage of infected cyst, sensation returns back to normal.
- *Salty taste*—when the cyst becomes secondarily infected and discharges into the mouth and when a sinus tract is present, then patient may complain of salty taste or a sinus tract is present.
- *Displacement of denture*—edentulous patients may seek treatment because of displacement of denture.
- *Tooth discoloration*—discoloration or loosening of tooth may prompt patients to visit the dentist.

Radiographic Features

- *View taken*—in the radiographic diagnosis of cysts of jaws, periapical and occlusal views are helpful. In addition, in case of large cysts, extraoral views provide an essential supplement to conventional intra-oral radiographs, which include lateral oblique, PA view for mandible, Waters' view and panoramic view (OPG).
- *Well defined radiolucency*—classically, radiographic appearance of a cyst is that of a well-defined round or oval areas of radiolucency circumscribed by a sharp radiopaque margin. Nevertheless, there is much variation to this standard pattern, which depends not only on the type of cyst but also on its location and the degree of bone destruction and expansion.

Other Diagnostic Features

- *Contrast study for cysts of the maxillary sinus*—in certain cases, like cysts in the maxillary sinus, use of contrast media, along with routine radiography is helpful in delineating the size and precise extent of the cyst. In this technique, the radio-opaque medium is injected into the maxillary sinus with an aspirating syringe. The mucosa is anesthetized and the needle is introduced into the maxillary sinus through the inferior meatus as for an aspiration biopsy of the sinus.
- *Ultrasonic diagnosis*—ultrasonography can be useful to give adequate information about the location and extent in depth of cysts. It is possible to make a diagnosis on the basis of the echo of an ultrasonic impulse and the margins of bone. It can also provide very interesting information for the diagnosis of conditions in the maxillary sinus.
- *Aspiration*—aspiration of a suspected cyst is a valuable diagnostic aid when doubt exists about the nature of lesion after careful clinical and radiographic examination. This examination is also helpful in distinguishing between maxillary sinus and maxillary cyst. Aspirate withdrawn from the cyst appears as straw colored fluid. Aspiration of air indicates penetration into the antrum. Failure to obtain air or fluid is an indication of a solid lesion.

Theories of Cyst Enlargement

The exact mechanism is not known, but it could be that the mechanism-governing enlargement of cysts of the jaws is the same irrespective of the type of cysts. The various steps involved in the formation of a cyst seem to be as follows:

- *Attraction of fluid*—the attraction of fluid into the cystic cavity.
- *Retention of fluid*—the retention of fluid into the cavity.

- **Raised hydrostatic pressure**—the production of raised internal hydrostatic pressure.
- **Resorption of surrounding bone**—the resorption of surrounding bone with an increase in the size of bone cavity.

Harries classified the theories of cyst enlargement in the following manner (**Fig. 13-1**)

- **Mural growth**
 - Peripheral cell division
 - Accumulation of cellular content
- **Hydrostatic enlargement**
 - Secretion
 - Transudation and exudation
 - Dialysis
- **Bone resorbing factor.**

Mural Growth

Peripheral cell division

- **Cell division of lining epithelium**—peripheral enlargement of cyst occurs due to active cell division of the lining epithelium. This is in response to an irritant stimulus. Cyst regression occurs following the removal of such stimulus.
- **Cons of theory**—the theory has been criticized on the basis that such regression would lead to an irregularly thickened inner surface because of the resistance of the surrounding bone. However, this ignores the possibility that the cyst wall is not only well supported by its fluid content but can also actively resorb bone sufficiently and rapidly to accommodate the expanding perimeter.

Accumulation of cellular content

- **Accumulation of mural squames**—Kramer has suggested that keratocyst enlarges by the increasing accumulation of mural squames which is the result of casting off the living epithelium. The characteristic finger-like projections of growth represents local areas of increased cell division. An alternative explanation for this elongation is that keratocyst although persistent in their growth are poor bone resorbers and simply extend preferentially along the less dense cancellous bone with little resorption and expansion of dense cortex.

Hydrostatic Enlargement

Growth of cyst occurs due to distension of the cystic wall by fluid that has accumulated by secretion, transudation and dialysis.

- **Secretion**—it occurs from goblet cells. Apart from the occasional goblet cells usually found in follicular cyst, there is little morphological evidence of intracystic secretion.

- **Transudation and exudation**—these have been proposed mainly for the enlargement of the follicular and periodontal cyst respectively. This conclusion was derived from an examination of the protein content and specific gravity of the cystic fluid. The presence of fibrin and cholesterol in periodontal and follicular cysts suggests that hemorrhage also contributed to the cystic fluid.
- **Dialysis**—the mean osmolality of the cystic fluid is 10 miliosmoles higher than that of serum. This gradient is attributed to the accumulation of the low molecular weight cells shed from the lining epithelium and maintained by inadequate lymphatic access to the cyst lumen, the consequence is net entry of fluid from the capsule capillaries into the cystic lumen.

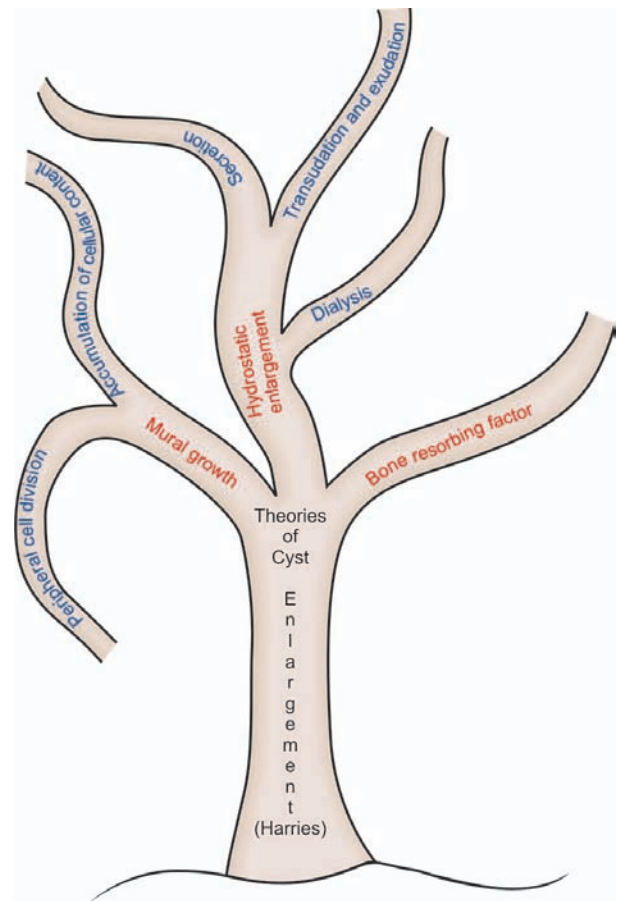


Fig. 13-1: Theories of cyst enlargement—diagrammatic representation

Bone Resorbing Factor

- **Prostaglandin**—vital cyst tissue in culture has been shown to release a potent bone resorbing factor, which is predominately a mixture of prostaglandin E_2 (PGE_2) and prostaglandin E_3 (PGE_3).
- **Mechanism**—the source of this resorbing factor appears to be the capsule and leukocyte content. Prostaglandin

release is reduced in some cysts when the epithelium is removed before culture but it is not clear whether the reduction results from the removal of an epithelial inductive effect or it merely reflects the loss of prostaglandin produced within the epithelium.

- *How prostaglandin production take place*—the mechanism of prostaglandin production is not known. One possibility is that production takes place in the capsule under the influence of epithelial proliferation, lysosomal phospholipase from fibroblasts and polymorphonuclear leukocytes; breaking down of phospholipid cell membrane to produce arachidonic acid which is converted by the ubiquitous enzyme prostaglandin synthetase to prostaglandin.

Classification

It is described in Tables 13-1 to 13-4.

Table 13-1: Classification of cysts by Robinson (1945)

From odontogenic tissues

Periodontal cyst

- Radicular or dental root apex type
- Lateral type
- Residual type
- Dentigerous cyst
- Primordial cyst

From non-dental tissues

- Median cyst
- Incisive canal cyst
- Globulomaxillary cyst

Table 13-2: Classification of cysts by WHO ICD-DA 1995 (excludes radicular cyst and mucus cyst)

Developmental odontogenic cyst

- Primordial (keratocyst)
- Gingival cyst
- Eruption cyst
- Dentigerous cyst (follicular)
- Gingival cyst of adults
- Lateral periodontal cyst
- Glandular odontogenic cyst
- Sialo-odontogenic cyst

Developmental non-odontogenic cyst

- Nasopalatine duct (incisive canal) cyst
- Globulomaxillary cyst (nowadays it does not include in cyst)
- Median palatal cyst (nowadays it does not exist)
- Cyst of palatine papilla

Other cyst of jaw

- Aneurysmal bone cyst (nowadays it is recognized as variant of giant cell tumor)
- Traumatic bone cyst (hemorrhagic bone cyst)
- Epithelial jaw cyst (no identifiable as odontogenic or non-odontogenic)

Other cyst of oral region not elsewhere classified

- Dermoid cyst
- Epidermoid cyst
- Gingival cyst of newborn
- Palatal cyst of newborn
- Nasoalveolar cyst (nasolabial)
- Lymphoepithelial cyst

Cyst of oral region unspecified

Table 13-3: Classification of cysts by Gorlin (1964)

Odontogenic cysts

- Dentigerous cyst
- Eruption cyst
- Gingival cyst of newborn infants
- Lateral periodontal and gingival cyst
- Keratinizing and calcifying cyst
- Radicular cyst
- Primordial cyst
- Multiple cysts of jaws and multiple cutaneous nevoid basal cell carcinoma and skeletal anomalies

Non-odontogenic and fissural cysts

- Globulomaxillary cyst (premaxilla maxillary cyst)
- Nasoalveolar (Nasolabial Klestadt's) cyst
- Nasopalatine (median anterior maxillary) cyst
- Median mandibular cyst
- Anterior lingual cyst
- Dermoid and epidermoid cyst
- Palatal cyst of newborn infants

Cysts of neck and oral floor and salivary gland

- Thyroglossal duct cyst
- Lymphoepithelial (bronchial cleft) cyst
- Oral cysts with gastric or intestinal epithelium
- Salivary gland cyst
- Mucocele and ranula

Pseudocyst

- Aneurysmal bone cyst
- Static (developmental latent) bone cyst
- Traumatic (hemorrhagic solitary) bone cyst

Table 13-4: Classification of cysts by Shear

Epithelial

Odontogenic

Developmental

- Dentigerous cyst (follicular)
- Eruption cyst
- Primordial cyst
- Gingival cyst of adults
- Lateral periodontal cyst
- Calcifying odontogenic cyst

Inflammatory cyst

- Radicular cyst
- Residual cyst
- Inflammatory collateral cyst
- Paradental cyst

Non-odontogenic

- Nasopalatine duct (incisive canal) cyst
- Median palatine, median alveolar and median mandibular cysts
- Globulomaxillary cyst
- Nasolabial cyst

Non-epithelial

- Simple bone cyst (traumatic solitary hemorrhagic bone cyst)
- Aneurysmal bone cyst

Cysts associated with maxillary antrum

- Benign mucosal cyst of the maxillary antrum
- Surgical ciliated cyst of maxilla

Cysts of the soft tissue of the mouth, face and neck

- Dermoid and epidermoid
- Branchial cleft cyst (lymphoepithelial)
- Thyroglossal duct cyst
- Anterior medial lingual cyst
- Oral cyst with gastric or intestinal epithelium
- Cystic hygroma
- Cysts of salivary glands
- Parasitic cyst, hydatid cyst, cysticercosis cellulosa

Epithelial Developmental Odontogenic Cyst

Dentigerous Cyst

It is also called as 'follicular cyst' or 'pericoronal cyst'. It is the most common type of odontogenic cyst which encloses the crown of the unerupted tooth by expansion of its follicle and is attached to the neck. Most of dentigerous cysts are discovered by radiographs, when these are taken because a tooth is missing or teeth are tilted or out of alignment.

Origin

- *Fluid accumulation*—it is formed due to fluid accumulation in the layer of reduced enamel epithelium or between it and the crown of unerupted teeth.
- *Proliferation of epithelium in dental follicle*—it may originate initially by proliferation and cystic transformation of islands of epithelium in the connective tissue wall of the dental follicle or even outside the follicle and this transformed epithelium then unite with the lining of follicle epithelium forming a solitary cystic cavity around tooth crown.



Fig. 13-2: Dentigerous cyst seen as swelling in canine region in maxillary area—intraoral view (Courtesy Dr Ashok L).

Clinical Features

- *Age and sex*—as it arises from the follicle of an unerupted tooth, it is usually found in children and adolescents with a higher incidence in 2nd and 3rd decades. It is slightly more common in males.
- *Site*—it is most commonly associated with mandibular 3rd molars and maxillary canines (**Fig. 13-2**) which are most commonly impacted. It may also be found enclosing a complex compound odontome or involving a supernumerary tooth.
- *Symptoms*—generally, it is painless but may be painful if it gets infected. When dentigerous cyst expands rapidly to compress sensory nerve, it produces pain which may be referred to other sites and described as headache.
- *Size*—they vary in size from a little more than the diameter of the involved crown to an expansion that causes progressive but painless enlargement of jaws and facial asymmetry (**Fig. 13-3**).
- *Teeth*—teeth adjacent to the developing cyst and involved teeth may get severely displaced and resorbed. There may be displacement of third molars to such an extent that it sometimes comes to lie compressed against the inferior border of the mandible.
- *Pathological fracture*—in some cases, pathological fracture can occur.
- *Expansion*—the most important feature of these lesions is its potential to expand. Cystic involvement of an unerupted third molar may result in hollowing out of the entire ramus extending up to the coronoid process and condyle as well as the body causing expansion of cortical plates (**Fig. 13-4**). In the case of a cyst associated with maxillary cuspids, expansion of the anterior maxilla often occurs and may superficially resemble acute sinusitis or cellulitis.
- *Associated disease*—bilateral cysts are found in association with basal cell nevus syndrome, cleidocranial dysplasia and a rare form of amelogenesis imperfecta.
- *Blue domed cyst*—when it contains blood, then it is called as 'blue domed cyst'.
- *Complication*—there are several potential complications besides the possibility of recurrence following incomplete surgical removal. These include:
 - *Ameloblastoma*—the development of an ameloblastoma either from the lining epithelium of the cyst or rests of odontogenic epithelium in the wall of the cyst, are called as mural ameloblastoma.

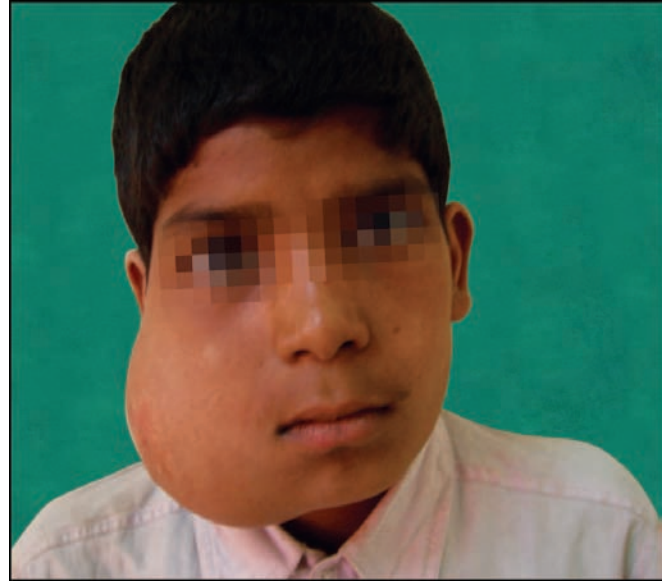


Fig. 13-3: Large dentigerous cyst causing facial asymmetry in patient (Courtesy Dr Parate).

- *Mucoepidermoid carcinoma*—the development of mucoepidermoid carcinoma, which is a malignancy of salivary glands, associated with the lining epithelium or dentigerous cyst which contain mucous secreting cell.



Fig. 13-4: Expansion in patient of dentigerous cyst (Courtesy Dr Bhaskar Patle).

Radiographic Features

- *Radiodensity*—well defined radiolucency usually associated with hyperostotic borders unless they are secondarily infected and are usually seen around an unerupted tooth (**Fig. 13-5**).
- *Internal structure*—usually, it is unilocular but sometimes it may appear multilocular, this image is caused by ridges in the bony wall and not by the presence of bony septa.



Fig. 13-5: Dentigerous cyst present with hyperostotic border.

- *Margins*—The bony margins are well defined and sharp (**Fig. 13-6**). In case of infection, margins may be ill defined.
- *Teeth*—It may envelop the crown symmetrically, but it may expand laterally from the crown. Cyst is usually



Fig. 13-6: Dentigerous cyst showing well defined border present in the mandible (Courtesy Dr Bhaskar Patle).

attached with the cemento-enamel junction. Associated tooth may be displaced in any direction. Usually direction of displacement is in apical region. Teeth may be displaced in the coronoid process, high up above the tooth-bearing region of maxilla.

- *Resorption of root*—the large cysts are always confined to the mandible. There may be resorption of roots of adjacent teeth.
- *Maxillary antrum*—the floor of the maxillary antrum may get displaced if cyst encroaches upon it.
- *Bilateral dentigerous cyst*—in some cases, there may be presence of dentigerous cyst on both side of mandible.
- *Multiple dentigerous cyst*—in some cases, we may find multiple dentigerous cyst present in one patient (**Fig. 13-7**).
- *CT features*—it shows expansive process with sclerotic border (**Figs 13.8 and 13.9**).
- *MRI features*—it shows homogenous high signal content.

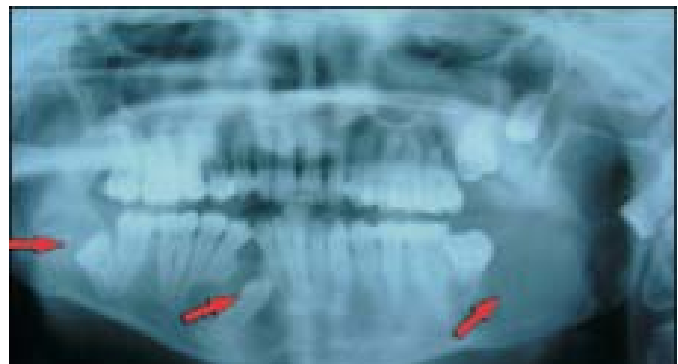


Fig. 13-7: Multiple dentigerous cyst present in mandible (arrow).

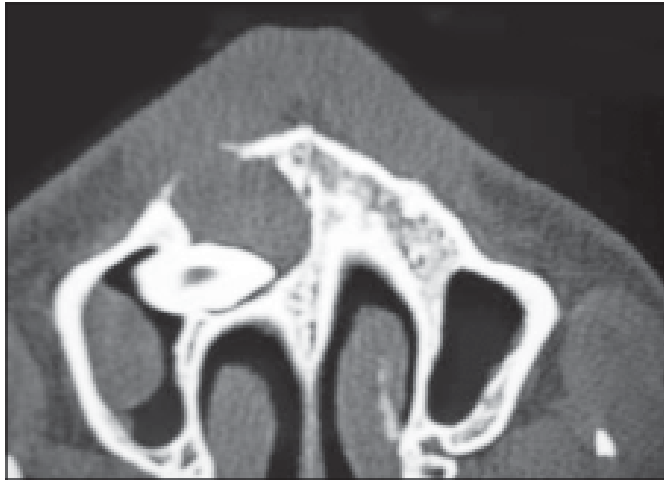


Fig. 13-8: CT scan of dentigerous cyst showing cyst associated with tooth.



Fig. 13-9: CT scan of dentigerous cyst present in posterior region of mandible (Courtesy Dr Iswar).

Radiographic Types

According to Thoma

- **Central variety**—in it, crown is enveloped symmetrically. In this instance, pressure is applied to the crown of the tooth and may push it away from its direction of eruption. In this way, the mandibular third molar may be found at the lower border of the mandible and in the ascending ramus and a maxillary canine in the sinus or as far as the floor of the orbit (Fig. 13-10). The maxillary incisors may be found below the floor of the nose.
- **Lateral type**—in it, dentigerous cyst is a radiographic appearance which results from dilation of the follicle on one aspect of the crown. This type is commonly seen when an impacted mandibular molar is partially erupted so that its superior aspect is exposed.
- **Circumferential type**—in it, the entire tooth appears to be enveloped by the cyst (Fig. 13-11). The entire enamel organ around the neck of the tooth becomes cystic often allowing the tooth to erupt through the cyst.



Fig. 13-10: Central variety of dentigerous cyst in which crown is enveloped by cyst.

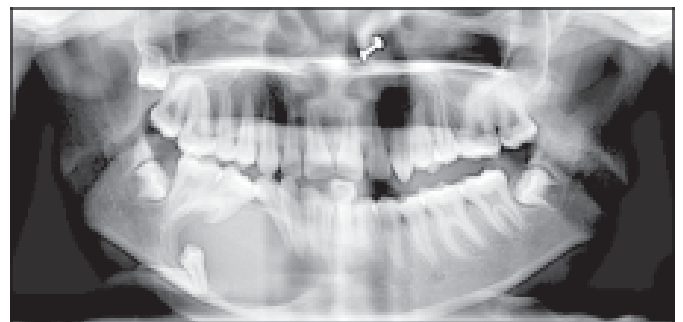


Fig. 13-11: Circumferential type of dentigerous cyst showing tooth enveloped by the cyst.

According to Mourshed

- **Class I**—dentigerous cyst associated with completely unerupted teeth
 - Dentigerous cyst associated with unerupted teeth, who failure to erupt is due to lack of space in the dental arch.
 - Dentigerous cyst associated with unerupted teeth, who failure to erupt is due to mal-positioning of the tooth germ.
 - Dentigerous cyst associated with unerupted supernumerary teeth.
- **Class II**—dentigerous cyst associated with partially erupted teeth.

Diagnosis

- **Clinical diagnosis**—expansive swelling in the posterior region of mandible will give clue to the diagnosis.
- **Radiological diagnosis**—well defined radiolucency associated with impacted teeth with hyperostotic border and well defined margin will diagnose the dentigerous cyst.

- *Laboratory diagnosis*—biopsy shows thin connective tissue wall with a thin layer of stratified squamous epithelium lining the lumen. Rete pegs formation is absent. It also shows Rushton bodies within the lining epithelium which are peculiar linear and often curved hyaline bodies. Fluid can be aspirated from the dentigerous cyst (Fig. 13-12).



Fig. 13-12: Aspirated fluid from dentigerous cyst (Courtesy Dr Shetty).

Differential Diagnosis

- *Ameloblastoma and ameloblastic fibroma*—they are multilocular and not associated with crown of an unerupted tooth. They will grow laterally away from the tooth in comparison to dentigerous cyst, which envelopes the tooth symmetrically and it is more common in premolar and molar area. There is internal structure present in ameloblastoma and in case of dentigerous cyst, it is most unlikely.
- *Adenomatoid odontogenic tumor*—they are rare and occur in the maxillary anterior region.
- *Calcifying odontogenic cyst*—it may occur as pericoronal radiolucency and may contain evidences of calcification.
- *Developmental primordial and follicular primordial cyst*—it can also be considered in the differential diagnosis. It occurs in close proximity to the crown of unerupted teeth and superimposition of its image causing cyst like radiolucency can appear as dentigerous cyst on the radiograph. However, in cases of follicular primordial cyst, the cystic lining surrounds the crown, whereas in dentigerous cyst, it is attached to the neck of tooth. The diagnosis of primordial cyst can be confirmed by taking a radiograph with different angulations.
- *Hyperplastic follicle*—the size of the normal follicle is 2 to 3 mm. If the follicular space exceeds 5 mm a dentigerous cyst is more likely the diagnosis. Region should be reexamined after 6 months to see any increase in size and effect on surrounding teeth.
- *Odontogenic keratocyst*—it does not expand the bone as severely as dentigerous cyst and also is less likely resorbs the tooth. It is usually attached more apically than the dentigerous cyst.
- *Radicular cyst at apex of primary teeth*—it may surround the crown of developing permanent teeth giving the false impression of dentigerous cyst. In this case, clinician should look for deep carious lesion or extensive restoration of the primary teeth which may help in the diagnosis of radicular cyst of primary teeth.

Management

- *Surgical*—smaller lesions can be surgically removed with little difficulty. The larger cyst involves surgical drainage and marsupialization. This procedure results in relief of pressure and gradual shrinking of the cystic lesion by peripheral opposition of new bone.
- *Decompression*—small acrylic button or short section of rubber is placed in preformed surgical opening in cyst which keeps the opening open and permits drainage. Recurrence is relatively uncommon unless there has been fragmentation of the cystic lining with remnants allowed to remain.
- *Orthodontic treatment*—in cases when you want to retain the tooth, orthodontic movement of teeth should be carried out.

Eruption Cyst

A specific type of cyst, which must be classified as a form of dentigerous cyst, is frequently associated with the erupting deciduous or permanent teeth in children.

An eruption cyst is in fact a dentigerous cyst occurring when a tooth is impeded in its eruption within the soft tissues overlying the bone, whereas the dentigerous cyst develops around the crown of an unerupted tooth which is lying in the bone. This cyst has often been termed as 'eruption hematoma'.

Pathogenesis

- *Dilation of tooth follicle*—it is essentially a dilation of the normal tooth follicle caused by accumulation of tissue fluid or blood. In 11% of cases, it occurs during the eruption of incisors and in 30% of cases, it occurs during eruption of canines and molars.

Clinical Features

- *Appearance*—clinically the lesion appears as a circumscribed, fluctuant, often translucent swelling of the alveolar ridge over the site of eruption of the tooth.
- *Age*—it is most commonly seen in children younger than 10 years of age.
- *Site*—it is most commonly seen first permanent molar and maxillary incisors.
- *Eruption hematoma*—when the circumscribed cystic cavity contains blood, the swelling appears purple or deep blue, hence it is termed as 'eruption hematoma'.

Radiological Features

- *Expansion of follicular space*—expansion of the normal follicular space of erupting tooth crown is seen.
- *Saucer shape excavation*—in some cases, there is saucer shaped excavation of bone projecting (Fig. 13-13) very slightly into the cavity.
- *Margin*—margin is well defined.

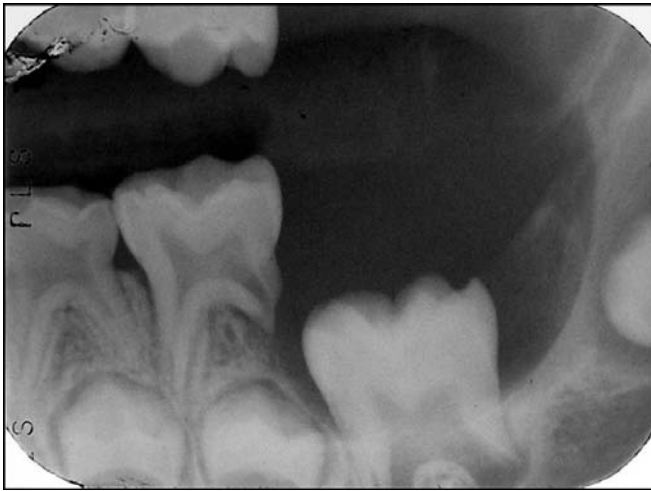


Fig. 13-13: Saucer shaped excavation of bone seen in eruption cyst (Courtesy Dr Ashok L).

Diagnosis

- *Clinical diagnosis*—circumscribed soft fluctuant translucent swelling around first permanent molar or maxillary incisor may give clue to the diagnosis.
- *Radiological diagnosis*—saucer shaped excavation of bone with well defined margin.
- *Laboratory diagnosis*—lamina propria shows variable inflammatory cell infiltrated.

Management

- *Excision of roof of cyst*—this is simple procedure to permit tooth to erupt. In some cases, cyst rupture spontaneously to facilitate tooth eruption.

Odontogenic Keratocyst

The keratocyst was probably first described by Mikulicz in 1876. According to Pindborg and Hansen, the designation keratocyst was used to describe any jaw cyst exhibiting keratinization in their lining which may occur in follicular, residual and very rarely in a radicular cyst.

11% of all jaw cysts are OKC. OKC is not a clinical diagnosis but a designation for a group of cysts of possibly diverse origins which have a number of highly characteristic microscopic and clinical features in common with highest recurrence rate of any of the odontogenic cyst.

In general, thin connective tissue wall and thin squamous type epithelial lining (4 to 8 cell thick) that contains keratin is either para (87%) or ortho (13%) and without rete pegs.

The basal layer is either columnar or cuboidal epithelium and its prickle cells are present and they are vacuolated. In some cases, bud-like proliferation from the basal layer into adjacent connective tissue wall or proliferation of islands of odontogenic epithelium that may be present in the wall giving rise to satellite microcysts which support the fact that OKCs have a high recurrence rate. 5% of dentigerous and radicular cysts are keratocysts and a variety of many primordial, gingival, lateral periodontal and residual cysts are of keratocyst variety.

According to Browne, odontogenic keratocyst is a histopathological term and should be restricted to its original sense and to describe the nature of the cyst. To avoid confusion between primordial cyst and keratocyst, Browne said that the term primordial cyst should be restricted to describe the origin and odontogenic keratocyst should be restricted to describe the nature of the cyst.

Origin of Cyst

- *Dental lamina*—it possesses marked growth potential or alternatively from proliferation of basal cells as 'basal cell hamartomas'.
- *Remnants of oral epithelium*—a residue or remnants of oral epithelium. The epithelium of the odontogenic keratocyst has been shown to be far more active than most odontogenic cysts as judged by their greater mitotic activity.

Clinical Features

- *Age*—odontogenic keratocyst occurs over a wide age range and cases have been recorded as early as the first decade and as late as the ninth with age group 4 to 84 years. Initiated early in life, during the period of tooth development with a peak incidence in 2nd and 3rd decades.
- *Sex*—it is found more frequently in males than in females and this sex predilection is more pronounced in blacks than in whites.
- *Site*—it is more common in mandible with a greater incidence at the angle and extending for varying distance into the ascending ramus and forward into the body (Fig. 13-14).
- *Symptoms*—asymptomatic unless they become secondarily infected, in which case patient complains of pain, soft tissue swelling and drainage. Occasionally, they experience paresthesia of the lower lip or teeth. Paresthesia can also occur, if there is a pathological fracture.



Fig. 13-14: Odontogenic keratocyst present as swelling in ramus area (Courtesy Dr Bhaskar Patle).

- **Teeth**—teeth may be displaced, if it expands through cancellous bone and the body of the mandible.
- **Signs**—the lesion can lead to pathologic fracture. As these cysts grow in anteroposterior direction, there is no obvious bony expansion seen (Fig. 13-15).
- **Aspiration**—on aspiration there is odorless, creamy or caseous content.
- **Syndrome associated**—multiple odontogenic keratocyst are found in Gorlin-Goltz syndrome, Marfan syndrome, Ehlers-Danlos syndrome and Noonan's syndrome.



Fig. 13-15: OKC intraoral view showing swelling in retromolar area (Courtesy Dr Tapasya Karamore).

Radiographic Features

- **Site**—more than 90% are seen posterior to the canines in the mandible and more than 50% at the angle of the mandible.
- **Characteristic**—40% have a characteristic suggestive of dentigerous cyst, 25% suggestive of primordial cyst, 25% lateral periodontal cyst and 10% of globulomaxillary cyst.
- **Internal structure**—undulating borders with cloudy interior appearances suggestive of multilocularity (Fig. 13-16).

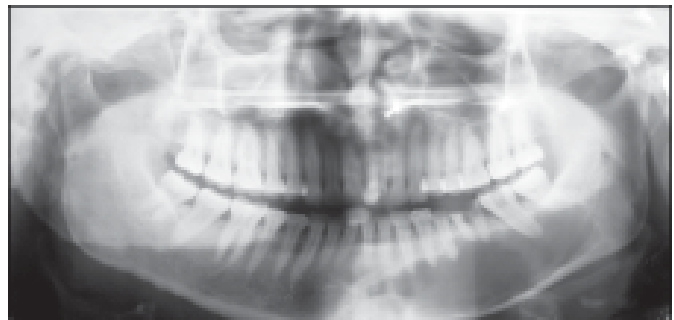


Fig. 13-16: OKC with undulating border and cloudy interior.

- **Size**—size varies and may be 5 cm or more in diameter. Maxillary lesions are smaller and rounder than those in the mandible.
- **Shape**—shape of cyst is usually oval extending along the body of the mandible with little mediolateral expansion.
- **Margins**—margins are hyperostotic.
- **Unilocular variety**—Majority of lesions are unilocular with smooth borders (Fig. 13-17) but some unilocular lesions are large with irregular borders. Radiolucency is usually hazy due to keratin filled cavity and it is surrounded by thin sclerotic rim due to reactive osteocytes.

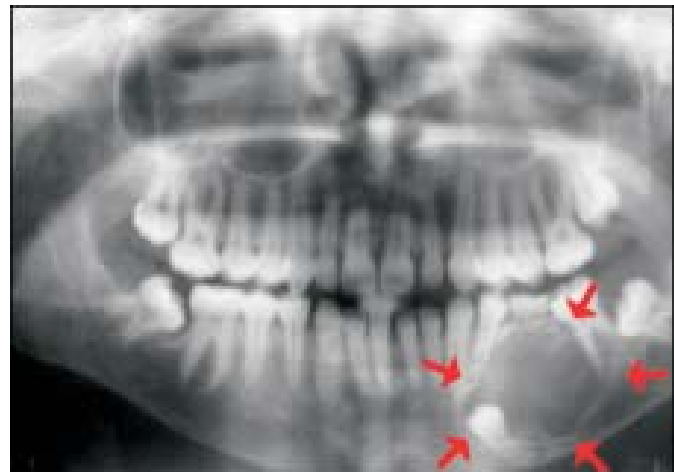


Fig. 13-17: Unilocular variety of OKC showing hyperostotic border (Courtesy Dr Bande).

- *Anteroposterior extension*—radiolucency extend along the medullary spaces in the anteroposterior direction (**Fig. 13-18**). There may be extensive involvement of the body and ascending ramus of the mandible with little or no bony expansion (**Fig. 13-19**).

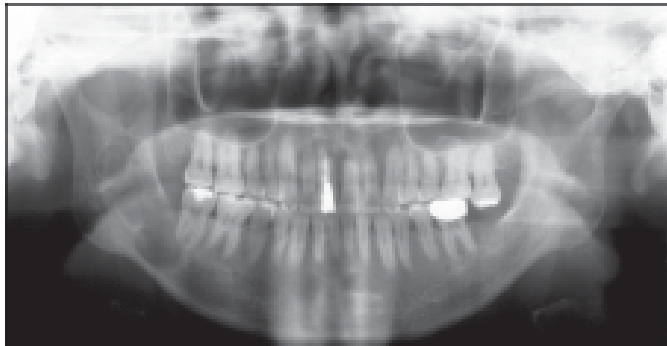


Fig. 13-18: Radiolucency extending in anteroposterior direction in OKC (Courtesy Dr Arijj).



Fig. 13-19: Extensive involvement of ramus and body of mandible in OKC.

- *Perforation of cortical plate*—in some cases it can perforate the buccal and lingual cortical plates of bone and involve the adjacent soft tissue.
- *Displacement of inferior alveolar canal*—downward displacement of the inferior alveolar canal and resorption of the lower cortical plate of the mandible may be seen as well as perforation of bone and a pathologic fracture may occasionally occur.
- *Teeth*—as the keratocyst enlarges it may produce deflection of unerupted teeth mostly in the region of the angle of the mandible and occasionally, on the ascending ramus and towards the orbital floor and in some cases, root resorption is also seen.
- *CT features*—it will demonstrate exact dimension of the radiolucency (**Fig. 13-20**).

Radiological Types of Keratocyst

- *Envelopment type*—it is referred to a variety of keratocyst which embraces an adjacent unerupted tooth.



Fig. 13-20: CT scan of OKC showing dimension of radiolucency (Courtesy Dr Iswar).

- *Replacement*—those which form in the place of normal teeth.
- *Extraneous*—those in the ascending ramus away from the teeth.
- *Collateral*—those adjacent to the root of teeth which are indistinguishable radiologically from the lateral periodontal cyst.

Diagnosis

- *Clinical diagnosis*—not so specific.
- *Radiological diagnosis*—radiolucency extending in anteroposterior direction with undulating border may be suggestive of OKC.
- *Laboratory diagnosis*—biopsy shows corrugated or wrinkled parakeratin surface. Prominent palisaded polarized basal cell layer often described as having a 'picket fence' or 'tombstone' appearance. Connective tissue shows daughter cysts or small satellite cysts.

Differential Diagnosis

It can be given in two ways:

Most likely

- *Ameloblastoma*—This usually occurs in older age. It is generally multilocular though a unilocular lesion may also occur. Mostly, it appears with paresthesia. As it is benign, it may show resorption of root with displacement. In cases of cyst, it shows amber colored fluid on aspiration.
- *Primordial cyst*—it is also common in third molar area. Absence of a tooth without a history of extraction favors primordial cyst. It also shows amber colored fluid on aspiration.

- *Residual cyst*—in cases of residual cyst, patient gives a history of extraction of tooth. Most of residual cysts are unilocular. A thin radio-opaque margin is common, although those infected will not have such a well defined margins. It shows amber colored fluid on aspiration.
- *Traumatic cyst*—the most characteristic radiographic feature of the cyst is the scalloped margins. Mostly, it appears unilocular. The lesion rarely causes cortical plate expansion but when this occurs it is mostly on buccal side. There may be history of trauma. In most cases, it is asymptomatic. Needle aspiration is usually non-productive. Only a few milliliter of straw colored or serosanguinous fluid can be withdrawn.

Less likely

- *Benign odontogenic tumor and cementifying or ossifying fibroma*—these are not common lesions.
- *Giant cell granuloma*—it is usually found in the anterior region of the jaw.
- *The giant cell lesion of hyperparathyroidism*—it can often be ruled out by studies on serum chemistry.
- *Tooth crypt*—the possibility of radiolucency of tooth crypt is less likely as calcification of the tooth starts in the 8th year of age. The halo has thin outer radio-opaque border which is continuous with the lamina dura in the area of the cemento enamel junction.
- *Odontogenic myxoma*—rarely myxoma must be considered in such circumstances because the tooth has failed to develop and may be seen as a cyst-like radiolucency.

Management

- *Enucleation*—enucleation of entire cyst with vigorous curettage of the cystic wall. Periodic post-treatment examination should be done.
- *Peripheral osteotomy*—peripheral osteotomy of bony cavity can be done to reduce chances of recurrence.
- *Chemical cauterization*—chemical cauterization of bony cavity with intraluminal injection of Carnoy's solution allow freeing the cyst from bony wall which in turn allow easier removal of cyst.
- *Decompression*—this is achieved with the help of polyethylene drainage tube kept in the bony cavity.

Recurrence

Rate of recurrence of odontogenic keratocyst is very high. There are number of reasons for it.

- *Satellite cyst*—occurrence of satellite cyst, which is a bud-like projection of basal cell layer into the connective tissue. Cystic lining may be retained during the enucleation procedure.
- *New cyst formation*—some instances of recurrence are likely because of new cyst formation rather than true recurrence.

- *Difficult in enucleation*—secondarily, its lining is very thin and fragile particularly when the cyst is large and therefore it is more difficult to enucleate than a cyst with thick wall. Portion of the lining may be left behind and constitute the origin of recurrence. Enucleation in one piece may be more difficult with cysts which have a scalloped and thin margins and this may explain the higher recurrence rate than those with smoother contour.
- *Intrinsic growth potential*—Toller suggested that there may be an intrinsic growth potential in the epithelial lining which may be responsible for a higher recurrence rate.
- *Proliferation of basal cell*—it may arise from proliferation of the basal cells of the oral mucosa particularly in the third molar area and ascending ramus of the mandible. It is referred that there is often a firm adhesion of the cyst to overlying mucosa and it is recommended that mucosa should be excised with them in an attempt to prevent possible recurrence from the residual basal cell proliferation.

Adenomatoid Odontogenic Cyst

Adenomatoid odontogenic cyst is cystic hamartoma arising from odontogenic epithelium. In some cases, proliferations fill all the lumen space mimicking a solid tumor.

Clinical Features

- *Location*—it is seen in anterior region of the maxilla.
- *Age and sex distribution*—it is common in young age group with predilection for women.
- *Symptoms*—it is usually asymptomatic. In some cases, expansion of jaw occur which is also associated with pain.
- *Size*—some of the lesion may enlarge up to size of 10 cm.
- *Signs*—expansion can be palpated and facial asymmetry can be present due to swelling.

Radiological Features

- *Appearance*—it appears as well demarcated unilocular radiolucency associated with impacted tooth (**Fig. 13-21**).
- *Calcification*—calcification can be seen. In some cases, this calcification will appear as small pebbles appearance.
- *Effect on surrounding structure*—separation of roots or displacement of an adjacent tooth occurs frequently. There is also cortical expansion and root resorption. The lesion may inhibit the eruption of involved tooth.

Differential Diagnosis

- *Dentigerous cyst*—it is seen in 2nd to 4th decade as compared to adenomatoid odontogenic cyst which is

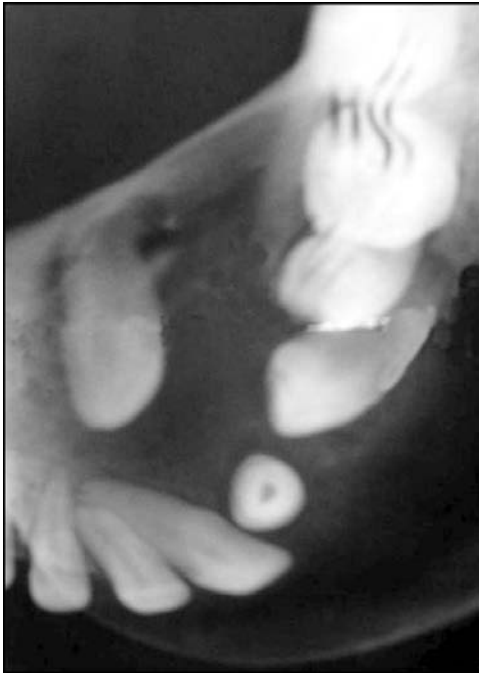


Fig. 13-21: Cystic radiolucency seen in anterior maxillary region suggestive of adenomatoid odontogenic cyst (Courtesy Dr Parate).

seen in young age. It is seen in posterior region as compared to adenomatoid odontogenic tumor which is seen in anterior region. Adenomatoid odontogenic tumor has tendency to surround more than just crown of the unerupted tooth.

- *Calcifying odontogenic cyst*—occurs in older age as compared to adenomatoid odontogenic tumor and mandibular premolar area is affected mostly.
- *Odontogenic fibroma or myxoma*—tennis racket pattern is seen.
- *Ameloblastic fibroma*—seen in premolar-molar region and has multilocular appearance.
- *Ameloblastic fibro-odontoma*—it is multilocular and radiopacities of enamel and dentin are seen inside the radiolucency as compared to adenomatoid odontogenic tumor where snowflakes are seen at the periphery.

Management

- *Exploration and enucleation*—AOC is treated by direct exploration and enucleation.
- *Marsupialization*—in some cases when lesion is associated with impacted tooth, marsupialization is done for eruption of tooth.

Lateral Periodontal Cyst

The lateral periodontal cyst is uncommon but a well recognized type of developmental odontogenic cyst. The designation lateral periodontal cyst is confined to that cyst,

which occurs as a result of inflammatory etiology and the diagnosis of collateral keratocyst has been excluded on clinical and histological ground.

Pathogenesis and Etiology

- *From dentigerous cyst*—initially origin is thought to be arising from dentigerous cyst which develops along the lateral surface of the crown. As the tooth erupts the cyst assumes a position in approximation to the lateral surface of the root.
- *Proliferation of cell rests of Malassez*—proliferation of cell rests of Malassez in the periodontal ligament may lead to lateral periodontal cyst. But stimulus which causes this proliferation is unknown.
- *Rests of dental lamina*—origin from proliferation and cystic transformation of rests of dental lamina (which is in post-functional state and therefore has only limited growth potential), that is in accordance with the usual small size of the cyst.
- *Post-functional dental lamina rests*—recent theory suggests that the lateral periodontal cyst and gingival cyst of adult share the common histogenesis from post-functional dental lamina rests. These two cysts represent basically the central or intra-osseous and peripheral or extra-osseous manifestations of the same lesion.

Types

- *Inflammatory*—it occurs near alveolar crest. Pocket content may irritate and stimulate rest of Malassez.
- *Developmental*—it is associated with developing tooth germ.

Clinical Features

- *Age and sex distribution*—the lateral periodontal cyst occurs chiefly in adults with an age range from 22 to 85 years with a mean age of 50 years. It shows a male predilection for occurrence.
- *Site*—the most frequent location of lateral periodontal cyst reported on lateral surface of the roots of vital teeth in mandibular canine and premolar region (**Fig. 13-22**) and is followed by the anterior region of the maxilla.
- *Symptoms*—gingival swelling may occur on the facial aspect and in such cases, it must be differentiated from the gingival cyst. In gingival cyst, the overlying mucosa is blue but in lateral periodontal cyst the overlying mucosa appears normal. When the cyst is located on the labial surface of the root, it appears as a slight obvious mass, overlying the mucosa.
- *Tooth*—the associated tooth is vital.
- *Infected cyst*—if the cyst becomes infected, it may resemble a lateral periodontal abscess.



Fig. 13-22: Ovoid radiolucency seen between roots suggestive of lateral periodontal cyst.

Radiographic Features

- **Shape**—the intra-bony lateral periodontal cyst is seen as a round or ovoid well defined radiolucency with hyperostotic borders.
- **Site**—it is usually found between the cervical margins and the apex of adjacent root surfaces and may or may not be in contact with root surfaces.
- **Size**—most of them are less than 1 cm in diameter except the botryoid variety which may larger and multilocular.

Variation of Lateral Periodontal Cyst

- **Botryoid odontogenic cyst**—this term was proposed by Waldron, which refers to a multilocular periodontal cyst. It resembles a cluster of lateral periodontal cysts exhibiting some difference. The lesion is multilocular with thin fibrous connective tissue septa. It is clear from numerous reports of recurrence that the botryoid odontogenic cyst requires careful excision.

Diagnosis

- **Clinical diagnosis**—normal color swelling seen in canine region.
- **Radiological features**—ovoid shaped radiolucency seen between roots of teeth with hyperostotic borders.
- **Laboratory diagnosis**—it is lined by a layer of stratified squamous epithelium with connective tissue wall. Cuboidal and columnar cells may be found compassing the lining. The lumen of the cyst shows focal thickened plaque of proliferating lining cell. These are especially prominent in botryoid cyst.

Differential Diagnosis

- **Lateral radicular cyst**—In radicular cyst, the lamina dura will not be intact and it is associated with pulpal infection and a non-vital tooth.
- **Lateral periodontal abscess**—it is very difficult to differentiate between abscess and cyst, but if it is less than 1.5 cm then it is considered as an abscess.
- **Lateral dentigerous cyst**—it is generally associated with an impacted tooth i.e. third molars and canines.
- **Residual cyst arising from the primary or permanent dentition**—there will be a history of extraction of the tooth.
- **Primordial cyst**—if primordial cyst arising from supernumerary tooth is superimposed on adjacent root surface, then it may be considered in differential diagnosis of lateral periodontal cyst. However, primordial cyst is more common at a younger age. So, to confirm the diagnosis of primordial cyst, radiographic examination from a different angulation should be carried out.
- **Globulomaxillary cyst**—it is seen in between maxillary lateral incisor and canine region common in young age and appears as a pear shaped radiolucency.
- **Median mandibular cyst**—it occurs in the midline of mandible.

Management

- The lateral periodontal cyst must be surgically removed if possible without extracting the associated tooth. If this cannot be accomplished, the tooth must be sacrificed. There is a tendency for recurrence for this type of cysts following its surgical excision.

Gingival Cyst of Adult

It is an uncommon cyst occurring either on free or attached gingiva. It is considered as soft tissue counterpart of lateral periodontal cyst.

Etiology and Pathogenesis

- **Proliferative epithelial tissue**—degenerative changes in proliferating epithelial tissue can occur which will result in gingival cyst of adults.
- **Remnants of dental lamina and enamel organ**—remnants of dental lamina, enamel organ or epithelial islands of periodontal membrane.
- **Trauma**—traumatic implantation of epithelium.
- **Post-functional rest of dental lamina**—it can also arise from post-functional rests of dental lamina.

Clinical Features

- **Age**—the gingival cyst may occur at any age, but it is most common in adults in the 5th and 6th decade of life.

- **Site**—the location of the lesion closely follows that of lateral periodontal cyst. It is more common in mandibular premolar and canine region with predilection for males.
- **Symptoms**—it is slowly enlarging, painless swelling, usually less than 1 cm in diameter and may occur in attached gingiva or the interdental papilla.
- **Appearance**—the surface may be smooth and the color may appear as that of normal gingiva or bluish (Fig. 13-23) and may appear red when it is blood filled as a result of recent trauma. Swelling is dome shaped.
- **Signs**—the lesions are soft and fluctuant and adjacent teeth are usually vital during surgical exploration.



Fig. 13-23: Bluish color swelling seen in premolar region due to gingival cyst of adults.

Radiographic Features

- **Superficial bone erosion**—there may be no radiological changes or only a faint round shadow indicative of superficial bone erosion (Fig. 13-24).



Fig. 13-24: Same patient (Fig. 13-23) showing slight erosion of underlying bone (Courtesy Dr Soni).

Diagnosis

- **Clinical diagnosis**—bluish color swelling of gingiva which is dome shaped will suspect gingival cyst of adults.
- **Radiological features**—bone erosion can be present.
- **Laboratory diagnosis**—biopsy has similar feature as that of lateral periodontal cyst.

Difference between Lateral Periodontal Cyst and Gingival Cyst of Adults

Although, it has been customary to consider these cysts as a distinctly different entity, their clinical appearance and behavior, morphologic and histochemical features and site of occurrence are so similar in appearance that they are in reality the intraosseous and extraosseous manifestations of the same pathosis. Bhaskar grouped the gingival and lateral periodontal cysts together as gingival cyst and considered that they both arise from extra-osseous odontogenic epithelium.

- **Location**—lateral periodontal cyst arises in the periodontium and located in the interproximal bone between the apex and the alveolar crest. Gingival cyst appears as a dome shaped swelling in the attached gingiva.
- **Radiological finding**—there is circumscribed radiolucency which occurs due to cup shaped depressions on the periosteal surface of cortical plates produced by enlargement of the gingival cysts. In case of lateral periodontal cyst there is round shaped radiolucency. The gingival cyst may certainly occur without bone involvement and may produce a gingival swelling. Most of the times, the swelling goes unnoticed. It is improbable though not impossible that a cyst originating in the gingival soft tissue could enlarge sufficiently to produce radiolucency by obvious bone erosion without producing any gingival swelling. In the case of a lesion, which has produced both gingival swelling and radiolucency, faint shadow (due to surface depression) indicates gingival cyst. Where the radiolucency is dark and sharply demarcated and a communication with the periodontium is indicated then the lesion is more likely to be lateral periodontal cyst that has eroded outward. These assumptions are based on radiological features and can be confirmed by surgical exploration when the lesion is being removed.

Management

- **Surgical excision**—surgical excision of the lesion in adults is usually recommended and the lesion does not tend to recur. A neoplastic potential has never been reported.

Palatal Cyst of Newborn (Epstein's Pearls, Bohn's Nodules)

Developmental cysts are common in newborn infant.

Pathogenesis

- *Entrapment of epithelium*—during the formation of secondary palate, small island of epithelium is entrapped below the surface along the median palatal raphe. Epithelium may be derived from minor salivary gland of palate.
- *Epstein's pearls*—these are cystic keratin filled nodules found along the midpalatine raphe probably derived from entrapped epithelial remnants along the line of fusion.
- *Bohn's nodules*—these are keratin filled cyst scattered over the palate most numerous along the junction of hard and soft palate and apparently derived from palatal salivary gland structure. The nodules are considered as remnants of mucus gland and are histologically different from Epstein's pearls.

Clinical Features

- *Appearance*—it is white or yellowish papule seen along the midline of palate.
- *Location*—it is present at junction of hard and soft palate.
- *Size*—it is usually 1 to 3 mm in diameter. Sometimes it may form clusters.

Management

- No treatment is required and cyst ruptures onto mucosal surface and eliminate their keratin content.

Dental Lamina Cyst

The cyst is apparently originated from remnants of dental lamina. They are multiple, occasionally solitary nodule on the alveolar ridge of newborn originating from remnants of the dental lamina (Fig. 13-25).

Clinical Features

- *Age*—these cysts are rarely seen after 3 months of age.
- *Site*—it is found on the crest of the maxillary and mandibular dental ridges.
- *Appearance*—clinically it appears as small whitish projection on the alveolar ridge of the jaws of infants giving mistaken appearance of a tooth (Fig. 13-26) at times appearing blanched due to internal pressure.
- *Size*—it is raised nodules, usually multiple, measuring a fraction of a millimeter to 2-3 mm in diameter.



Fig. 13-25: Dental lamina cyst showing elevation on alveolar ridge (Courtesy Dr Bhaskar Patle).



Fig. 13-26: Dental lamina cyst giving appearance of raised nodule on alveolar ridge (Courtesy Dr Bhaskar Patle).

Diagnosis

- *Clinical diagnosis*—whitish soft tissue elevation on alveolar ridge in infant will diagnose this condition.
- *Laboratory diagnosis*—there is thin epithelial lining and shows lumen filled with desquamated keratin, occasionally containing inflammatory cells.

Management

- No treatment is required as these lesions almost invariably disappear by rupturing onto the surface of the mucosa or through disruption by erupting teeth.

Calcifying Epithelium Odontogenic Cyst (CEOC)

It is also called as 'keratinizing and calcifying odontogenic cyst, dentinogenic ghost cell tumor, calcifying ghost cell odontogenic cyst' and 'Gorlin's cyst' as it was first described by Gorlin in 1962. The lesion is unusual in that it has some

features suggestive of cyst but also has many characteristic of a solid neoplasm.

Types

Clinical

- *Central or intraosseous variety*—it is also called as intraosseous odontogenic cyst. Cyst occurs centrally within the bone.
- *Peripheral or extraosseous variety*—it may occur peripherally in the soft tissue overlying the tooth bearing area.
- *Associated with odontogenic tumor*—this variety associated with odontogenic tumor.

Prattorius

He divided this variety into three types:

- Simple unicystic type.
- Unicystic odontome producing type.
- Unicystic ameloblastomatous proliferating type.

Clinical Features

- *Age and sex distribution*—most of the cases are diagnosed in 2nd and 3rd decade of life. It is slightly more prevalent in women.
- *Site*—3/4th of the lesions occur centrally, with about equal in both jaws and 75% occurring anterior to the first molar.
- *Symptoms*—it is slow growing, painless, non-tender swelling of the jaws. Occasionally, some patients may complain of pain.
- *Signs*—in some cases, cortical plate over the expanding lesion may be destroyed and cystic mass may be palpable with patients reporting of discharge. Aspiration yields viscous, granular, yellow fluid.
- *Teeth*—adjacent teeth may be displaced.

Radiographic Features

- *Radiodensity*—the central lesion may appear as a cyst like radiolucency.
- *Margins*—it presents with variable margins which may be quite smooth with a well defined outline or irregular in shape with poorly defined borders.
- *Teeth*—it is not unusual to find these associated with unerupted teeth and in some cases as pericoronal radiolucency. Roots of adjacent teeth may show resorption.
- *Internal structure*—it may contain small foci of calcified material that are only microscopically apparent. In some cases, it is completely radiolucent while in other cases an increasing amount of calcified material may be seen as white flecks. It may be unilocular or multilocular. In some cases, calcified material may occupy most of the lesions.

Diagnosis

- *Clinical diagnosis*—not specific.
- *Radiological features*—cystic lesion with calcification seen inside the bony cavity (**Fig. 13-27**) may give clue to the diagnosis.
- *Laboratory diagnosis*—focal areas of stellate reticulum and ghost cell may be present as well as sparse amount of dentinoids may be seen. Ghost cells are pale eosinophilic swollen epithelial cells that have lost its nucleus and nuclear membrane.

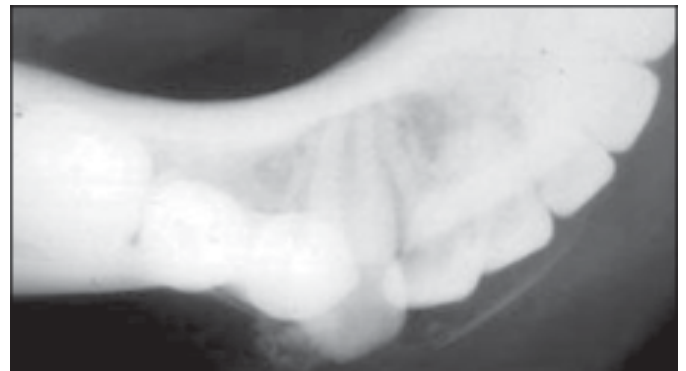


Fig. 13-27: Calcifying epithelial odontogenic cyst showing some degree of calcification.

Differential Diagnosis

- *Fibrous dysplasia (initial stage)*—it appears as mottled or smoky defined border on radiographs. It has poorly defined borders. It is more common in the maxilla.
- *Partially calcified odontoma*—it appears within the capsule.
- *Adenomatoid odontogenic tumor (AOT)*—in intermediate stage of development of AOT, radiographically it appears like CEOC.
- *Ossifying fibroma (initial stage)*—the fibro osseous lesions are likely to be situated in more inferior position in the mandible. It may show root resorption. Histologically, it shows 'Chinese letter' shaped islands of bone or calcification distributed throughout the connective tissue.
- *Odontogenic fibroma*—histologically, it shows odontogenic tissue like cementum.
- *Cementoblastoma*—the radiographic image is well defined and attached to the root of tooth.

Management

- *Surgery*—enucleation and curettage should be done.

Glandular Odontogenic Cyst

It is also called as '*Sialo odontogenic cyst*', or '*mucoepidermoid cyst*'. It is developmental odontogenic cyst which also shows salivary or glandular features.

The most descriptive term of the lesion is probably mucoepidermoid odontogenic cyst because of the presence

of both secretory elements and stratified squamous epithelium.

Clinical Features

- *Age*—it is more commonly occur in middle aged adults.
- *Site*—it is more commonly seen in mandible with slight predilection for anterior region.
- *Size*—size is usually less than 1 cm in diameter.
- *Symptoms*—small lesions are asymptomatic but large lesions may associate with pain and paresthesia.
- *Sign*—expansion may be present in large lesion.

Radiological Features

- *Appearance*—it is a unilocular or multilocular radiolucency with either smooth or scalloped margins (**Fig. 13-28**).
- *Margin*—margin of the lesion is well defined with sclerotic margin.

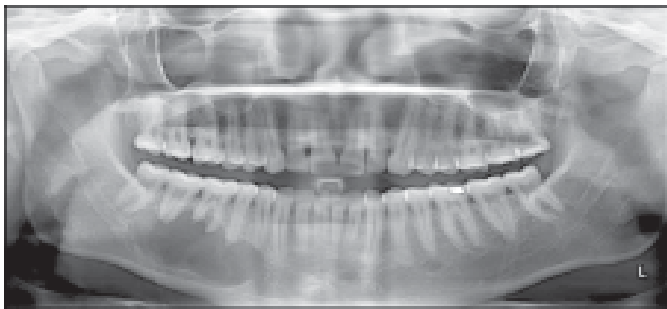


Fig. 13-28: Glandular odontogenic cyst present with scalloped margin in posterior region of mandible.

Diagnosis

- *Clinical diagnosis*—not specific.
- *Radiological features*—multilocular appearance with scalloped margin may suspect this cyst.
- *Laboratory diagnosis*—histologically, it shows cystic space lined by non-keratinized epithelium. The mucus and cylindrical cells form an integral part of the epithelial component with mucinous material within the cystic space.

Management

- *Surgery*—enucleation or curettage should be performed in this case. Recurrence is common in this type of cyst.

Inflammatory Cysts

Radicular Cyst

It is also called as 'apical periodontal cyst', 'periapical cyst', or 'dental root end cyst'. It is a common sequelae in progressive

changes associated with bacterial invasion and death of the dental pulp. It most commonly occurs at the apices of teeth.

The radicular cyst is classified as an inflammatory cyst because this process is thought to initiate the growth of the epithelial component.

Pathogenesis and Origin

- *Periradicular inflammatory changes*—periradicular inflammatory changes cause the epithelium to proliferate. As the epithelium grows into a mass of cells, the center loses the source of nutrition from the periapical tissue. These changes produce necrosis in the center and a cavity is formed and cyst is created.
- *Abscess cavity formation*—another theory is that an abscess cavity is formed in the connective tissue and is lined with proliferating epithelial tissue.
- *Origin of epithelium*—origin of cyst may occur from cell rests of Malassez which are remnants of Hertwig's root sheath and is a product of the odontogenic epithelial layer.
- *Other sources of epithelium origin*—the epithelium may be derived in some cases from respiratory epithelium of the maxillary sinus, when the periapical lesion communicates with the sinus wall, oral epithelium proliferates apically from the periodontal pocket and oral epithelium forms a fistulous tract.

Clinical Features

- *Age*—peak frequency occurs in the 3rd decade and there are large numbers of cases in the 4th and 5th decades after which there is a gradual decline. Although dental caries is more common in deciduous teeth in the first decade but radicular cysts are not often found in deciduous teeth may be because teeth tend to drain more readily than the permanent teeth and the antigenic stimuli which evoke the changes leading to the formation of radicular cyst may be different.
- *Sex*—male predominance, as females are less likely to neglect their maxillary anterior teeth and males are more likely to sustain trauma to their maxillary teeth. It shows a higher frequency of occurrence in whites.
- *Site*—maxillary anterior are more commonly affected. Also in addition to caries, maxillary teeth are more prone to traumatic injuries which lead to pulp death.
- *Symptoms*—it represents an asymptomatic phase in periapical inflammatory process following death of the dental pulp. It is associated with non-vital tooth (**Fig. 13-30**).
- *Signs*—it rarely causes non-tender expansion of the overlying cortical bone. Intraoral and extraoral swelling may be visible in some cases (**Fig. 13-29**).



Fig. 13-29: Radicular cyst involving upper premolar region showing extraoral swelling (Courtesy Dr Bhaskar Patle).



Fig. 13-30: Intraoral swelling in association with non-vital central incisor in case of radicular cyst (Courtesy Dr Bande).

- **Teeth**—the associated tooth is not sensitive to percussion.
- **Consistency**—swelling may be bony hard or crepitations may be present as bone is thinned or it may be rubbery or fluctuate, if the bone is completely destroyed.
- **Complication**—ameloblastoma, epidermoid carcinoma and mucoepidermoid carcinoma may arise in the epithelial lining of periapical cyst.

Radiographic Features

- **Shape**—it appears as a rounded or pear shaped radiolucency at the apex of non-sensitive tooth or with non-vital tooth (**Fig. 13-31**).
- **Site**—it appears on the mesial or distal surface of a tooth root, at the opening of an accessory canal or infrequently

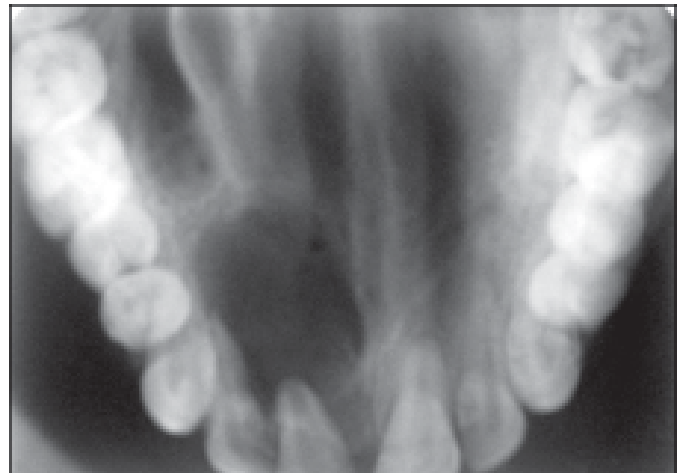


Fig. 13-31: Radicular cyst, occlusal view seen as pear shaped radiolucency.

in a deep periodontal pocket. In some cases, radicular cyst can be associated with deciduous teeth (**Fig. 13-32**).

- **Size**—radiolucency is more than 1.5 cms in diameter but usually less than 3 cms in diameter. It has got well-defined outline with thin hyperostotic borders.

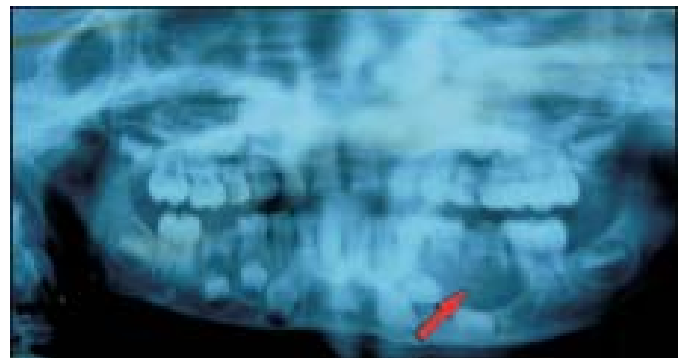


Fig. 13-32: Radicular cyst associated with deciduous teeth.

- **Margins**—in uncomplicated cases, margins are smooth, corticated and the cortex is usually well defined, well etched and continuous, except in some cases, where, there may be window formation. There is also thin white line surrounding the margins of the bone cavity. This thin layer of cortical bone is almost always present unless suppuration supervenes in the cyst.
- **Resorption of root**—radicular cysts of long duration may cause resorption of roots.
- **Adjacent teeth**—adjacent teeth are usually displaced and rarely resorbed (**Figs 13-34 and 13-35**).
- **Maxillary antrum**—if it involves maxillary area then displacement of antrum can occur. Due to distal inclination of the root (**Fig. 13-33**), cyst which arises from lateral incisor may invaginate the antrum.
- **Cortical plate**—the outer cortical plates of the maxilla or mandible may expand in a curved or circular shape.

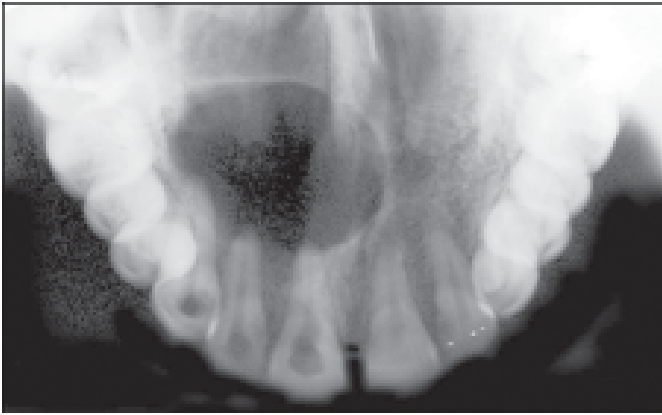


Fig. 13-33: Radicular cyst present in the maxillary anterior region.



Fig. 13-34: Radicular cyst causing displacement of anterior teeth.



Fig. 13-35: Radicular cyst seen associated with two teeth.

- *Alveolar nerve canal*—cyst may displace the mandibular alveolar nerve canal in inferior direction.
- *CT finding*—CT image shows expansive process with sclerotic border.
- *MRI finding*—T1 weighted image shows oval expansive process with homogenous high signal. There may be rim enhancement.

Diagnosis

- *Clinical diagnosis*—swelling associated with non-vital teeth may give clue to the diagnosis.
- *Radiological diagnosis*—well defined radiolucency with size more than 1.5 cm is associated with non-vital teeth with loss of lamina dura will confirm the diagnosis of radicular cyst.
- *Laboratory diagnosis*—cyst is lined by stratified squamous epithelium. Hyaline bodies or Rushton bodies often found in great numbers in the epithelium of apical periodontal cyst. Collection of cholesterol cleft with associated multinucleated giant cells is found in the wall of the lesion.

Differential Diagnosis

- *Periapical granuloma*—if the radiolucency is smaller than 1.5 cm then it is most likely to be a granuloma. Cysts show a straw colored fluid on aspiration. Cyst is more apt to have thin hyperostotic borders than granuloma. Morse and coworkers described chair side method for differentiating cysts and granuloma. This method involves the use of alkaline copper tartarate in cystic fluid aspirate. A consistent color difference related to difference in protein content was observed for cyst and granuloma. Aspirate of root canal fluid from patients with cyst showed an intense albumin pattern and definite pattern in globular zones on polyacrylamide gel electrophoresis and fluid associated with periapical granuloma on the other hand shows only a faint-to-moderate pattern in the albumin zone.
- *Periapical scar*—it can be eliminated on the basis of history and location.
- *Surgical defect*—it can be diagnosed on the basis of history.
- *Lateral periodontal cyst*—radicular cyst originates from maxillary lateral incisor and positioned between the roots of the lateral incisor and canine and may get confused with lateral periodontal cyst. In such case, vitality of the tooth should be checked. Tooth in relation with the lateral periodontal cyst is vital and with the radicular cyst it is non-vital.
- *Periapical cementoma (early stages)*—in cementoma, the tooth is vital whereas the tooth with granuloma or cyst has a non-vital pulp. Periapical cementoma mostly involves the incisors.

- *Traumatic bone cyst*—the pulp of associated tooth is usually vital. 90% of traumatic cysts occur in mandible, i.e. in molar and premolar area whereas the periapical cyst has no predilection for occurrence.
- *Periodontal abscess*—it usually shows moderate to severe crestal bone loss. Mostly the tooth is vital.
- *Mandibular infected buccal cyst*—it is more common in young patients. First molar is frequently involved. Pulp is usually vital. The lamina dura around the tooth is intact.
- *Benign tumor*—presence of septum inside the cavity is suggestive of benign tumor.

Management

- *Root canal treatment*—it is the treatment of choice as in many cases, radicular cyst resolve after root canal treatment. The reason behind it is that as drainage is established, the inflammatory process subsides and the fibroblast start producing collagen. The pressure of proliferating collagen reduces the blood supply to the epithelium lining and causes it to degenerate. Macrophages remove the degenerating epithelial tissue.
- *Extraction*—if the lesion fails to resolve, extraction of associated tooth is carried out.
- *Enucleation and marsupialization*—enucleation or marsupialization of a larger lesion is done.

Residual Cyst

It is a cyst that either remained as such in the jaw when its associated tooth was removed or was formed in residual epithelium of cell rests from a periodontal ligament of the lost tooth. Low grade inflammation of parent cyst might predispose formation of residual cyst.

Clinical Features

- *Age and sex*—cyst is common in patients older than 20 years with an average age of about 52 years. It is twice more common in male than female.
- *Site*—higher incidence in the maxilla. Mostly found on alveolar process (**Fig. 13-36**) or body of the tooth bearing area with some cysts described in the lower ramus of the mandible.
- *Symptoms*—it is asymptomatic with a previous history of pain in the tooth.
- *Size*—it is seldom more than 5 to 10 mm in diameter.

Radiographic Appearance

- *Pre-extraction radiograph*—pre-extraction radiographs show tooth with an evidence of deep caries or fractured tooth adequate for pulp involvement and/or an associated cyst.



Fig. 13-36: Swelling seen on alveolar ridge due to residual cyst (Courtesy Dr Tapasya Karamore).

- *Shape*—it is round or ovoid radiolucency that is usually well circumscribed (**Fig. 13-37**).
- *Margins*—thin radio-opaque margins are common with unilocular appearance although the infected cyst will not have such well defined margins.
- *Appearance*—due to resistance, cyst enlarges in elliptical shape with hyperostotic borders resulting from minimal pressure of expanding cyst.

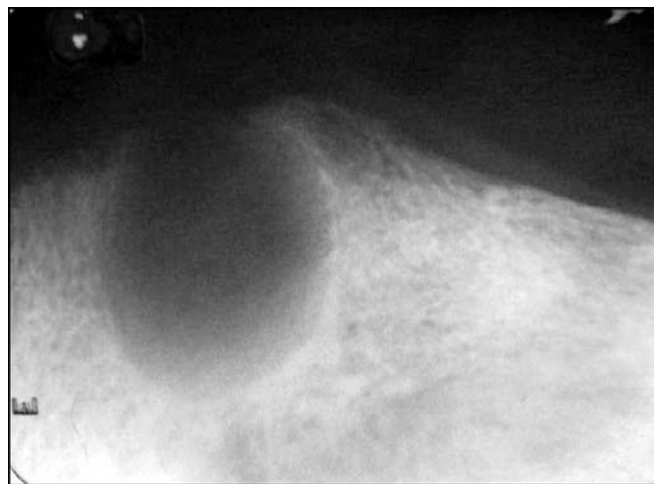


Fig. 13-37: Residual cyst seen as well defined radiolucency (Courtesy Dr Tapasya Karamore).

Differential Diagnosis

- *Primordial cyst*—in it, tooth is missing without a history of extraction. It is more common in mandibular posterior teeth and at a younger age. On aspiration, it shows straw colored fluid.
- *Keratocyst*—it is more common in mandibular posterior area and at an early age. It may appear multilocular. The size of the cyst is larger when compared to residual cyst.

- *Traumatic cyst*—usually in the mandible, in apical region (if it is associated with tooth) it shows scalloped outline. It is common at younger age. Aspiration is non-productive.
- *Ameloblastoma* (in initial stages/unilocular)—usually the lesion appears larger compared to the residual cyst. There is no history of extraction of the tooth.

Diagnosis

- *Clinical diagnosis*—not specific.
- *Radiological diagnosis*—well defined radiolucency with previous history of endodontic treatment and pre-extraction radiograph showing radiolucency.

Management

- *Enucleation*—If the cyst is not large and patient's age and health will tolerate the insult, the cyst wall should be completely enucleated. The extent of repair of the defect will depend on the size of the cyst and health of the patient.
- *Marsupialization*—in cases where the surgical procedure must be as atraumatic as possible, marsupialization or decompression may be used.
- *Excision*—complete excision and replacement with autogenously bone graft or single segment of bone fixed in it.

Inflammatory Collateral Cysts

They arise in the periodontium on the lateral aspect of an erupted tooth as a result of inflammatory process in the periodontal pocket. They arise by proliferation of cell rests of Malassez in the lateral periodontium. They are rare as drainage occurs more readily from the lateral periodontium which is closely associated with gingival crevice.

Paradental Cyst

It was first described by Craig in 1976. It is a cyst of inflammatory origin occurring on the lateral aspect of the root of partially erupted mandibular third molar with an associated history of pericoronitis.

Origin

- *Cell rests of Malassez*—it can be the origin, but argument is that if rests of Malassez were responsible then the lesion should be equally distributed around the root surface.
- *Reduced enamel epithelium*—second theory is that it originates from the reduced enamel epithelium as the presence of reduced enamel epithelium over the enamel projection might be the source and could explain the common location of the cyst.

Clinical Features

- *Age and sex*—it occurs between 10 to 39 years of age. It is most common in the third decade of life. It shows predilection for males.
- *Site*—usually associated with the third molar on the buccal surface (**Fig. 13-38**) and covers the bifurcation. It may occur bilaterally.
- *Signs*—the involved tooth is vital.



Fig. 13-38: Radiolucency seen distal to third molar in case of paradental cyst.

Radiographic Features

- *Radiodensity*—there is well demarcated radiolucency occurring distal to the partially erupted tooth but there was often buccal superimposition.
- *Intact periodontal ligament space*—the radiolucency sometimes extends apically but an intact periodontal ligament space provided the evidence that the lesion did not originate at the apex.

Diagnosis

- *Clinical diagnosis*—not so specific.
- *Radiological features*—radiolucency distal to third molar with intact periodontal ligament space.
- *Laboratory diagnosis*—cyst is lined by proliferating, non-keratinized squamous epithelium of varying thickness. The fibrous capsule is the seat of an intense chronic or mixed inflammatory cell infiltrate.

Management

- *Enucleation*—the lesion is treated by surgical enucleation.

Mandibular Buccal Infected Cyst or Buccal Bifurcation Cyst

It is an inflammatory cyst occurring on the buccal surface of mandibular molars in young children. The reason for

occurrence of this cyst is that inflammation response may occur in surrounding follicular tissue which stimulated cyst formation.

Clinical Features

- **Age**—a younger age group than the paradental cyst.
- **Site**—it affects the mandibular first and second molars more commonly rather than the wisdom teeth. The cyst is always situated on the buccal surface of the mandibular molar most frequently the first permanent molar after their partial or complete eruption (**Fig. 13-39**).
- **Symptoms**—there may be discomfort, pain, tenderness and rarely suppuration. Swelling, particularly if inflamed, is the clinical feature most likely to induce the patient to seek advice. Facial swelling may follow and this may be inflamed.
- **Signs**—pulp is vital in this cyst.
- **Teeth**—the associated tooth is usually tilted, so that the apices are adjacent to the lingual cortex, a feature of which is demonstrable in occlusal radiographs.
- **Size**—the size of cyst varies and may extend beyond the limit of the involved tooth and impinge upon and displace the crypt of the adjacent unerupted tooth.
- **Expansion**—the cyst extension in buccal direction is variable, but frequently the outer bony cortex is lost.
- **Pocket**—periodontal probing will show pocket formation on buccal side of first molar.

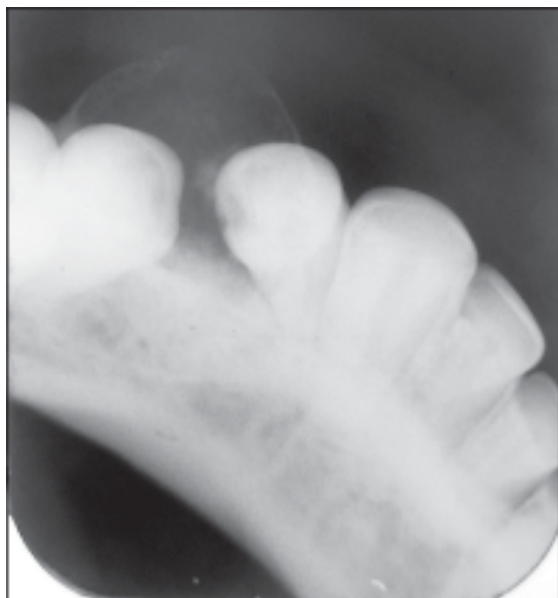


Fig. 13-39: Radiolucency associated with mandibular first molar with extension upto premolar with buccal expansion suggestive of buccal bifurcation cyst (*Courtesy Dr Parate*).

Radiographic Features

- **Periosteum**—with involvement of the periosteum, new bone may be laid down either as a single linear band or

laminated, if there are two or more layers. Sometimes, the new bone may be homogeneous.

- **Site**—the cysts will appear on the buccal aspects of the affected molars.
- **Bone**—usually there is involvement of the bone in the furcation and the entire interradicular bone may be lost.
- **Margins**—the inferior margins of the cyst is concave and rarely, the cyst may extend to the inferior border of the mandible but not leading to any external deformity.

Diagnosis

- **Clinical diagnosis**—the diagnostic features are the young age of the patient, the mandibular molar sites, buccal periostitis, usually vital pulp.
- **Radiological features**—intact lamina dura with buccal expansion visible on radiograph.
- **Laboratory diagnosis**—not so specific.

Management

- **Enucleation**—it is generally agreed that enucleation of the cyst without removal of the associated tooth is the treatment of choice.

Suppurating Cyst

When pyogenic organisms are present in sufficient number to produce changes, suppuration results. The thin white line which represents the cortex of the cyst becomes less dense and thinner and may be entirely lost (**Fig. 13-40**). In rare cases there is slight irregularity of the walls of the cyst, with some decalcification extending into the bone to a short distance. In the place of thin white cortex, there is sometimes a broad band of sclerosed bone which is less dense but wider than a normal cortex and presents a granular appearance.

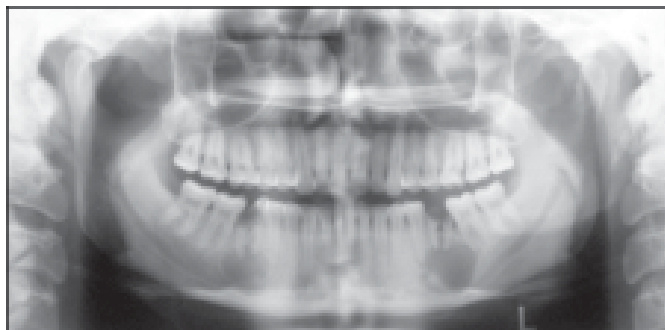


Fig. 13-40: Loss of thin line of cyst due to suppuration on left side of radicular cyst in molar area (*Courtesy Dr Tapasya Karamore*).

Healing Cyst

A cyst that is caused by an infected tooth often heals after removal of that tooth. One of earliest change is the gradual loss of the cortical layer of bone which lines most of the

cysts. At the same time, if the cyst becomes infected, the bone in the immediate vicinity of the walls becomes radiolucent as a result of hyperemia.

After the period of time, there is gradual lessening of radiolucency of the cavity and a return of the surrounding bone to a normal density (Fig. 13-41). The dark shadow of the cavity is replaced by a grey one and it is apparent that the cavity is becoming smaller. A large cyst may fail to heal completely, resulting in a smaller cyst. This is due to the retention of some part of the epithelial wall of the cyst, which has continued its activity.

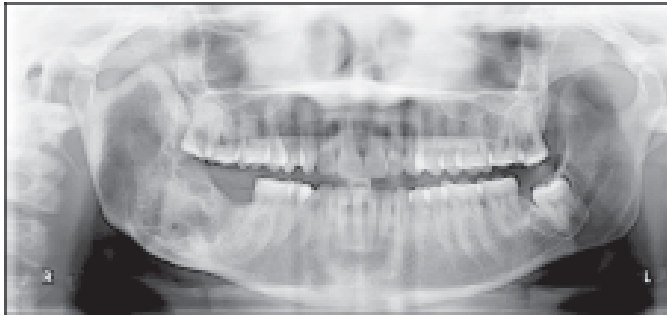


Fig. 13-41: Cyst is in healing stage. Note increased density of the bone in the mandibular right ramus region.

Non-odontogenic Cysts

Nasopalatine Cyst

It is also called as 'nasopalatine duct cyst', 'incisive canal cyst', 'median anterior maxillary cyst' or 'vestigial cyst'. It is the most common non-odontogenic cyst in the oral cavity. It is generally agreed that the nasopalatine duct cyst is an entity. It may occur within the nasopalatine canal or in the soft tissue at the opening of the canal where it is called as *cyst of the palatine papilla*. They are found to form in the canal or at the oral terminus of the canal.

Etiopathogenesis

- **Development**—it is developmental in origin and arises in the incisive canal when embryonic epithelial remnants of the nasopalatine duct undergo proliferation and cystic transformation. Some workers believe that it is derived from the epithelium included in the lines of fusion of embryonic facial processes.
- **Trauma**—in the form of direct blow to the incisive canal or indirectly from mastication, particularly when ill-fitting dentures are involved, have been suggested. But if this is true, the cyst would likely to be found more often and there would not be a male predilection.
- **Bacterial infection**—either from the nasal cavity or from the oral cavity, stimulates to the epithelial remnants to proliferate has been suggested as a cause. However, since

an open connection with the oral cavity or the nasal cavity is extremely rare, an evidence for the bacteria is lacking.

- **Retention phenomenon**—blocked duct of mucus glands causes an accumulation of secretion would be responsible for the cystic expansion.

Clinical Features

- **Age and sex distribution**—most cases discovered in the 4th and 6th decades and it is also frequently found in edentulous patients. It is three times higher in males than in females and found equally in blacks and whites.
- **Onset**—there is a small well defined swelling just posterior to the palatine papilla.
- **Pain**—sometime it may become infected, producing pain. Burning sensation and numbness may be experienced due to pressure on the nasopalatine nerve.
- **Salty taste**—patients complain of salty taste in mouth produced by small sinus or remnant of nasopalatine duct that permits cystic fluid to drain into oral cavity.
- **Fluctuant**—swelling is fluctuant and bluish if it is near the surface.
- **Palpation**—it is opened by a tiny fistula on or near the palatine papilla. In such cases, a tiny drop of watery fluid or pus may be elicited by pressure in this area.
- **Surface**—deeper cysts are covered by normal mucosa, unless it is ulcerated.
- **Expansion**—if cyst expands, it may penetrate the labial plate and produce a swelling below the maxillary labial frenum (Fig. 13-42).
- **Teeth**—roots of central incisors diverge. It may bulge into the nasal cavity and distort nasal septum.
- **Cyst of palatine papillae**—Sometimes, cyst formed in palatine papilla will be evident as an elevation or a soft round swelling of palatine papilla which extends posteriorly along the midline of the palate. It occurs anterior to the incisive foramen below the periosteum and do not enter invades the underlying bone.



Fig. 13-42: Nasopalatine duct cyst showing swelling in palate (Courtesy Dr Shetty).

- **Complication**—in some cases, it is reported that it may lead to malignancy of maxilla.

Radiological Features

- **Site**—typical cyst like radiolucency is superimposed with the apices of the central incisors. If cyst is formed in one of the branches of the canal, image will be displaced on one side of the midline.
- **Heart shape radiolucency**—image of radiopaque anterior nasal spine may superimpose over the dark cystic cavity giving it a heart shape (**Fig. 13-43**) or shape of inverted tear drop.
- **Shape**—in many cases, cyst is situated symmetrically in the midline. It may be rounded, oval or irregularly shaped.
- **Size**—size of the cyst is variable but usually from 6 mm to several centimeters in diameter.
- **Paired cyst**—two separated cysts may develop in two canals and cause paired cysts like radiolucency.
- **Erosion of bone**—cyst in canal cavity also erodes the bone posterior to the canal and creates impression of midpalatal cyst.
- **Teeth**—adjacent teeth usually show distal displacement but they are rarely resorbed. Divergence of central incisor roots and external root resorption is common.
- **Cyst of palatine papillae**—the cyst in the palatine papillae is not usually apparent because it does not cause enough pressure on the bone to cause discernible bone resorption but it can produce some bony erosions.



Fig. 13-43: Incisive canal cyst giving heart shaped radiolucency in incisor region.

Differential Diagnosis

- **Incisive fossa**—The shape of the fossa varies from round to oval to triangular to diamond to funnel shaped. Radiolucency in the area less than 6 mm wide should be considered as a normal fossa in the absence of associated

symptoms. Incisive fossa sharply defines at the lateral margins in contrast to the cyst, which has a well defined boundary on all margins. Aspiration will help to distinguish between cyst and incisive fossa.

- **Radicular cyst**—Pulp is non-vital with loss of lamina dura in the radicular cyst.
- **Dentigerous cyst with mesiodens**—Radiographic evidence of association with supernumerary teeth will establish the diagnosis of dentigerous cyst.
- **Median palatine cyst**—radiolucent lesion is behind the incisive canal in premolar-molar area.
- **Primordial cyst from supernumerary teeth**—more common in the posterior teeth.

Diagnosis

- **Clinical diagnosis**—swelling posterior to palatine papilla may give clue to the diagnosis
- **Radiological diagnosis**—heart shaped radiolucency in maxillary anterior region is typical of nasopalatine duct cyst.

Management

- **Surgical enucleation**—its removal is not indicated unless there are clinical symptoms. Removal is indicated in edentulous patients before dentures are introduced. Intraoral approach should be taken for the surgical enucleation.

Nasolabial Cyst

It is also called as 'nasopalveolar cyst' or 'Klestadt's cyst'. It is a soft tissue cyst, which involves the bone secondarily.

Pathogenesis

- **Fissural cyst**—some state that it is a fissural cyst arising from epithelial rests which are entrapped in the fusion lines of medial nasal, lateral nasal process and maxillary process.
- **Misplaced epithelium of nasolacrimal duct**—others state that it is actually a merging of mesenchymal processes and not a fusion per se, so there is no epithelial entrapment in the naso-optic fissure. They state that the location of nasopalveolar cyst strongly argues in favor of its development from the embryonic nasolacrimal duct that initially lies on the surface.

Clinical Features

- **Age and sex distribution**—it can occur at any age group with peak in 3rd and 4th decade of life. Women are more commonly affected in the ratio of 3:1.
- **Site**—it is unilateral but may be bilateral.

- **Symptoms**—it may cause pain and difficulty in breathing through the nose. There is swelling of the nasolabial fold and nose. In some cases, swelling may rupture spontaneously and it may drain into oral cavity.
- **Signs**—swelling is fluctuant on palpation. There is flaring of ala and distortion of nostril and fullness of upper lip below the nasal vestibule. It may bulge into the nasal cavity and cause some obstruction. Infection may drain into the nasal cavity.
- **Histopathological features**—it may contain goblet cells.

Radiographic Features

- **Site**—it is located above the apices of incisor in alveolar bone in maxillary region.
- **Radiodensity**—it is radiolucent lesion. Cyst causes an erosion of the underlying bone by virtue of its presence and pressure.
- **Margins**—usual outline of inferior border of nasal fossa is distorted resulting in posterior convergence of the margins.
- **Shape**—the actual shape and position of the cyst can be demonstrated by aspirating the typical cyst fluid and replacing it with radiocontrast material. A tangential view then demonstrates the *kidney shaped lesion* below the nasal fossa and above the apices of the incisors.

Differential Diagnosis

- **Acute dentoalveolar abscess**—in this case, tooth is tender on percussion. Radiographically, cystic radiolucencies are seen in case of nasoalveolar cyst.
- **Nasal furuncle**—no radiographic evidence of the lesion.
- **Mucus extravasation cyst**—no radiographic sign are seen.
- **Cystic salivary adenoma**—same as above.

Diagnosis

- **Clinical diagnosis**—difficulty in breathing from nose, flaring of ala of nose and distortion of nostril will diagnose nasolabial cyst.
- **Radiological diagnosis**—not specific.

Management

- **Intraoral surgical approach**—it should be excised using an intraoral approach. Some portion of nasal mucosa should also be removed for complete removal of cyst. There is no tendency to recur.
- **Endoscopic marsupialization**—in this case, transnasal approach is taken.

Anterior Alveolar Cyst

It is also called as '*median alveolar cyst*'. This cyst arises in the intermaxillary suture anterior to the site of anterior

palatine cyst. It arises near the labial surface of the premaxilla between the two incisors. There may be swelling at the site.

Radiologically, it occupies the position more close to the shadow of the crowns of the incisors than the root. It appears as area of radiolucency in the midline. The borders may be sharply defined or not. There may or may not be a thin layer of cortical bone at the periphery of the cavity. In some cases, when the cyst comes in contact, there is symmetrical resorption of the roots of teeth.

Nonepithelial Cysts

Idiopathic Bone Cavity

It is also known as '*solitary bone cyst*', '*hemorrhagic bone cyst*', '*extravasation cyst*', '*traumatic bone cyst*', '*simple bone cyst*', '*unicameral cyst*' and '*idiopathic bone*'. It is an unusual lesion which occurs with considerable frequency in the jaws as well as in other bones of the skeleton.

The traumatic bone cyst is a misnomer, since these intra-bony cavities are not lined by epithelium. It may results from trauma induced intra-medullary hematoma with subsequently result in bone resorption and cavitations during hematoma resolution.

Pathogenesis

- **Intramedullary hemorrhage**—it originates from intra-medullary hemorrhage following traumatic injury. Hemorrhage occurring within the medullary space of bone after trauma heals by organization of the clot and eventual formation of connective tissue and formation of new bone.
- **Traumatic injury theory**—according to the traumatic injury theory, however, it is suggested that after traumatic injury to an area of spongy bone containing hemopoietic marrow enclosed by layer of dense cortical bone, there is failure of organization of blood clot and for some unexplained reasons, subsequent degeneration of the clot that eventually produce an empty cavity within the bone.
- **Degeneration of clot**—in the development of the lesion, the trabeculae of bone in the involved area become necrotic after degeneration of the clot and bone marrow, although some viable marrow tissue must persist to initiate resorption of the involved trabeculae.
- **Infiltrating edema**—the lesion then appears to increase in size by steady expansion produced by a progressive infiltrating edema on the basis of restriction of venous drainage. This expansion tends to cease when the cyst-like lesion reaches the cortical layer of the bone, so that expansion of the involved bone is not a common finding in the solitary bone cyst.

- *Other causes of origin of cyst*—it may originate from bone tumors that have undergone cystic degeneration, as a result of faulty calcium metabolism such as that induced by parathyroid disease. It may originate from necrosis of faulty marrow due to ischemia, end result of low grade chronic infection and as a result of osteoclastic activity resulting from disturbed circulation caused by trauma thereby creating an unequal balance of osteoclastic activity and repair of bone.

Clinical Features

- *Age and sex*—the traumatic bone cyst occurs most frequently in young persons at an age of 6 to 20 years with a male predominance as they are exposed to traumatic injury most frequently than females with the ratio being 3:2.
- *Site*—it is usually found in mandible anywhere from the symphysis to the ramus, but about one-third are found in the maxilla, usually in the anterior region.
- *Symptoms*—it is asymptomatic in most cases but occasionally, there may be evidence of pain and tenderness.
- *Signs*—cortical swelling or slight tooth movement are not the usual finding and the teeth are vital.
- *Aspiration*—needle aspiration is actually unproductive and if it is productive, it contains either a small amount of straw colored fluid shed off necrotic blood clot and fragment of fibrous connective tissue.

Radiographic Features

- *Site*—they may be found in dentulous as well as edentulous arch.
- *Appearance*—it appears as a radiolucent lesion with a spectrum of well defined to moderately defined borders.
- *Margins*—most cases are unilocular with a fairly regular border. There is evidence of hyperostotic borders around the entire lesion but occasionally, such border is lacking. Most characteristic radiographic feature of this cyst is scalloped superior or occlusal margins where it extends between the roots of the teeth.
- *Size*—some cysts may be only a centimeter in diameter while others may be so large that they involve most of the molar area of the body of the mandible as well as part of the ramus.
- *Teeth*—they may be superimposed with the root or 'scallop' superiorly between the roots. Occasionally, lamina dura of the tooth may be absent and even less frequently the root may show resorption.
- *Alveolar bone*—it rarely causes cortical expansion but if it occurs, it is mostly buccal (**Fig. 13-44**). Surface is smooth, the cortical plates are not disrupted and pathological fracture does not result. The cavity occupies the position in the body of the jaw, but it may extend to involve the alveolar process.

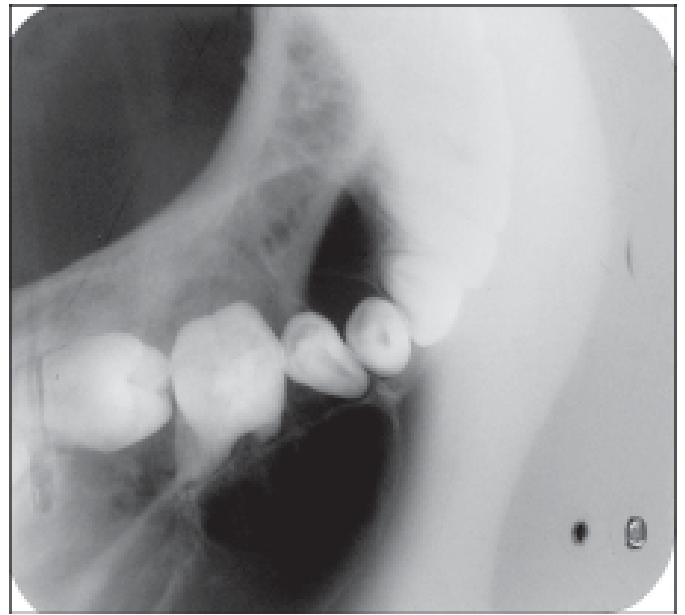


Fig. 13-44: Idiopathic bone cavity with thinned cortex and expansion of bone (Courtesy Dr Tapasya Karamore).

Diagnosis

- *Clinical diagnosis*—it is very difficult to make clinical diagnosis.
- *Radiological diagnosis*—well defined radiolucency with vital tooth with history of trauma will give clue to diagnosis.
- *Laboratory diagnosis*—aspiration is non-productive. Biopsy shows fragments of fibrin with enmeshed red cells may be seen. Hemorrhage and hemosiderin fragments are usually present and scattered small cells are often found.

Differential Diagnosis

- *Radicular cyst*—in radicular cyst, tooth is usually non-vital. Further, all the true cysts tend to have a more rounded appearance.
- *Central giant cell granuloma*—it usually shows evidence of internal bony septa where traumatic bone generally lacks. It is more common in mandibular anterior region.
- *Ameloblastoma and odontogenic myxoma*—are usually multilocular.
- *Lesions of eosinophilic granuloma*—these lesions are not well corticated as that of traumatic bone cyst.
- *Fibrous dysplasia*—not so corticated.

Management

- *Surgical exploration*—simple surgical exploration to establish the diagnosis. When the correct diagnosis is determined, enucleation and curettage are carried out.
- *Intralesional steroid injection*—this can yield limited success in some cases of traumatic bone cyst.

Cysts of the Maxillary Sinus

It is discussed in Chapter 27: Disorders of Maxillary Sinus.

Cysts of Soft Tissues of the Face and Mouth

Epidermoid Cyst

Epidermoid cyst is a simple cyst which is filled with cyst fluid or keratin and no other specialized structure. Epidermoid cyst is lined by epidermis, but contains no appendages. Epidermoid cyst is also called as *infundibular cyst* as most of cysts are derived from follicular infundibulum.

Pathogenesis

- *Trauma*—implantation keratinizing epidermoid cyst may occur in other parts of the mouth as a result of trauma. This type of cyst is also called as *epidermal inclusion cyst*.
- *Infundibular epithelium*—there is non-neoplastic proliferation of infundibular epithelium from healing process of localized inflammation of hair follicle.

Clinical Features

- *Age and sex distribution*—it is more common in 2nd and 3rd decade of life. Males are more commonly affected as compared to female.
- *Location*—these are most common on head, neck, face and back of the patient.
- *Appearance*—it is nodular lesion (Fig. 13-45) which may be associated with inflammation.



Fig. 13-45: Epidermoid cyst showing nodular appearance on the face of patient (Courtesy Dr Bande).

- *Fluctuation test*—it is positive.
- *Syndrome associated*—it is associated with Gardner's syndrome.

Diagnosis

- *Clinical diagnosis*—nodular lesion on face with positive fluctuant test will give clue to the diagnosis.

Management

- *Conservative surgical excision*—it is treated by conservative surgical excision.

Dermoid Cyst

It may occur as developmental anomalies and about 1 to 2% occurs in oral cavity. It contains sebaceous material as well as keratin. If lumen contains elements such as bone, muscles or teeth from various germinal layer is called as '*teratoma*'. Dermoid cyst is benign cystic form of teratoma. Dermoid cyst is lined by epidermis and skin appendages are present in the fibrous wall.

Types

- *Median variety*
 - Supramylohyoid variety
 - Inframyllohyoid variety
- *Lateral variety*
 - Supramylohyoid variety
 - Inframyllohyoid variety

Clinical Features

- *Age and sex*—it occurs at any time from birth to adolescence and it is small in infancy and large in adolescence. Mainly, it is apparent between 12 to 25 years of age and occurrence is equal in both sexes.
- *Site*—midline of the floor of mouth is the commonest location of these cysts which may cause a swelling in the midline of the neck or sometimes, it may be lateral. On the face, it can be seen on the forehead (Fig. 13-46).
- *Symptoms*—swelling is slow and painless. In the case of supramylohyoid variety, swelling may displace the tongue and it may interfere with breathing, speaking, closing the mouth and eating.
- *Size*—the size may vary up to several centimeters in diameter.
- *Palpation*—swelling is soft to firm, rubbery or cheesy consistency and sharply delineated.
- *Aspiration*—it contains straw colored fluid.
- *Transillumination test*—it is usually negative.
- *Fluctuation test*—it is generally positive.
- *Color and surface*—if superficial, it is yellow to white and surface is smooth and non-ulcerated until traumatized.



Fig. 13-46: Dermoid cyst presenting as well defined swelling on the forehead of patient.

- *Movement with tongue*—it does not move with protrusion of the tongue or deglutition.
- *Double chin appearance*—inframylaryoid variety also causes swelling in the submental area which gives rise to a 'double chin' appearance.

Radiographic Appearance

- *CT scan*—CT scan may be useful for the detection of dermoid cyst.
- *Contrast study*—in some cases, if you want to see the extent of the cyst, then we have to remove some content of the cyst to enable introduction of an opaque substance such as lipiodol, which is chemical combination of iodine and poppy seed oil. After this, radiograph is made from every position necessary to enable opaque material find its way by gravity to each portion of the cyst.

Differential Diagnosis

- *Ranula*—it is not in midline and appears bluish. Transillumination test is positive.
- *Unilateral or bilateral blockage of Wharton's duct*—in this case patient experience pain during meals.
- *Thyroglossal duct cyst*—it lifts when the patient swallows or protrudes the tongue.
- *Cellulitis*—the swelling is diffuse and widespread.
- *Submandibular lymph nodes swelling*—it is solid.

Diagnosis

- *Clinical diagnosis*—midline location is typical feature of this disease.
- *Radiological diagnosis*—contrast study will diagnose this lesion.

Management

- *Surgical excision*—it is the treatment of choice. Supramylaryoid variety should be excised through intraoral approach and inframylaryoid variety should be excised taking extraoral approach.

Lymphoepithelial Cyst

Its location in the neck, parotid gland and other intraoral areas will be dealt separately. Lymphoepithelial cyst of neck is called as 'branchial cleft cyst'.

Pathogenesis

- *From cervical sinus*—caudal overgrowth of second arch covers the grooves of second, third and fourth branchial arch. These grooves lost contact with outside and from the ectoderm line cervical sinus. Failure of these cervical sinus results in formation of branchial cyst.
- *From glandular epithelium*—it may originate due to cystic transformation of glandular epithelium entrapped within the oral lymphoid aggregates during embryogenesis. Epithelium in lymphoepithelial cyst might be derived from the ductal epithelium of salivary glands.
- *Parotid gland lymphoepithelial cyst*—cystic changes occur in parotid gland epithelium which becomes entrapped in upper cervical lymph nodes during embryonic life.
- *Oral lymphoepithelial cyst*—lymphoid tissue is found in oral cavity in the area of palatine tonsil, lingual tonsil and pharyngeal adenoids. Lymphoid aggregates may also occur in the floor of mouth, ventral surface of tongue and soft palate.

Clinical Features

Lymphoepithelial cyst of the neck

- *Age*—occurs at all ages with a fairly equal distribution from first to sixth decade.
- *Site*—location is superficially in the neck, close to the angle of the mandible, anterior to the sternocleidomastoid muscle.
- *Size*—the neck lesions vary in size from small to very large (about 10 cm in diameter).
- *Symptoms*—there is swelling, which may be progressive or intermittent and pain may also be a feature.
- *Sign*—it is soft, fluctuant swelling. Some of the cyst may form mucoid discharge on to the skin through the opening.

Lymphoepithelial cyst of parotid gland

- *Age*—the age ranges from 16 to 69 years with a male to female ratio of 3:1.
- *Symptoms*—there is a nodule in the parotid gland.
- *Signs*—it is most commonly associated with HIV infection.

Oral lymphoepithelial cyst

- **Age**—the age range is 15-65 years with a male predominance.
- **Site**—it affects mainly the floor of mouth and the tongue.
- **Appearance**—the cyst appears as a non-ulcerated freely movable mass which has been present for a period ranging from 1 month to a year.
- **Symptoms**—patients may complain of swelling and discharge.
- **Size**—size may vary from few millimeters to 2 cm.
- **Signs**—fairly mobile, superficial soft fluctuant, sharply delineated swelling.
- **Shape and color**—the usual observation was of round or oval swelling of the oral mucosa of normal color but when large swelling they were yellow or white in color.

Management

- **Surgical excision**—surgical removal of the cyst should be carried out. There are no chances of recurrence.

Thyroglossal Duct Cyst

It is also called as 'thyroglossal tract cyst'. The analogue of the median lobe of the thyroid gland develops at about the fourth week of the intrauterine life from a site at the base of tongue which is recognized later as the foramen caecum. A hollow epithelial stalk known as the thyroglossal duct extends caudally and passes ventrally to the hyoid bone to the ventral aspect of the thyroid cartilage where it joins the developing lateral lobe. The thyroglossal duct disintegrates by about tenth week of intrauterine life, but cyst may form from residue of the duct at any point along its line of descent.

Pathogenesis

- **Origin**—it develops from remnant of thyroglossal duct.
- **Stimulation**—it is stimulated by inflammatory conditions which lead to reactive hyperplasia of the lymphoid tissue near remnant of thyroglossal duct.
- **Accumulation**—this reactive hyperplasia block the thyroglossal duct, resulting in accumulation of secretion and formation of cyst.

Clinical Features

- **Age and sex distribution**—it is slightly more common in women and occurs commonly in the first, second and third decades of life.
- **Site**—most commonly located in midline in the area of hyoid bone and when they occur in the mouth, they are either in the floor or at the foramen caecum.
- **Symptoms**—it is painless and movable swelling. Pain may occur if the cyst is infected. If they are located high in the tract, they may cause dyspnea.

- **Shape**—it is spherical or oval with a long axis along the thyroglossal tract.
- **Size**—the cyst varies in size from 0.5 to 5 cm in diameter.
- **Signs**—they lift when the patient swallows or protrudes the tongue. It is fluctuant and tender swelling. Consistency is often firm or hard depending upon the tension of fluid within the cyst.
- **Movement with deglutition**—cyst moves with deglutition as swelling is attached to hyoid bone by fibrous tissue. It also moves with protrusion of tongue.
- **Fluctuation and transillumination**—it is usually positive.
- **Complication**—complication like fistula formation, recurrent infection can occur.

Differential Diagnosis

- **Subhyoid bursal cyst**—it moves with deglutition but not with protrusion of tongue.
- **Sublingual dermoid**—it does not move with protrusion of the tongue.

Management

- **Sistrunk operation**—it involves the removal of a 1 cm block of tissue surrounding the duct and the duct should be traced down to the pyramidal lobe of thyroid gland and to the foramen caecum at the base of tongue.

Anterior Median Lingual Cyst

It is also called as 'intralingual cyst of foregut origin'. It is a rare lesion and is commonly found in children. It can be present since birth and recurrence is common. It is a fluctuant swelling in anterior half of the tongue. There may be difficulty in feeding.

Oral Cyst with Gastric or Intestinal Epithelium

It is also called as 'oral alimentary tract cyst'.

Pathogenesis

- **Location of primitive stomach**—Gorlin pointed out that in the 3-4 mm embryo, the undifferentiated primitive stomach lies in the mid-neck region, not far from analogue of the tongue. Gastric mucosa has been shown to occur in the esophagus of 7.8% of infant and in 51% of these, heterotrophic tissues were located in the upper third.
- **Fusion of endodermal and ectodermal epithelia**—it is suggested that in the sublingual region of the oral cavity and in the region of apex and dorsum of tongue, the ectodermal and endodermal epithelia fuse and this will explain presence of heterotrophic gastric or intestinal mucosa.

Clinical Features

- *Age and sex*—most cases occur in infants and young children between the ages of 2 to 11 years. The male to female ratio is 3:1.
- *Site*—common location is in the anterior part of the tongue, floor of mouth and submandibular gland. The cyst may be enclosed entirely within the tongue or floor of mouth or may communicate with the surface.
- *Appearance*—swelling may be seen in tongue.

Diagnosis

- *Clinical diagnosis*—not specific.
- *Laboratory diagnosis*—the cyst may be lined partly by stratified squamous epithelium and partly by gastric or intestinal mucosa.

Management

- *Surgical excision*—surgical excision should be carried out. Recurrence is rare but has been reported.

Cystic Hygroma

It is a developmental abnormality in which there is progressive dilatation of lymphatic channels.

Clinical Features

- *Age*—it is often present at birth and most cases are diagnosed before the age of 2 years.
- *Site*—it frequently involves the neck and face, although it can occur anywhere in the body.
- *Symptoms*—those that involve facial tissue produce a swelling often painless and usually compressible.
- *Signs*—the overlying skin may be blue and the swelling transilluminates. There may be a history of gradual or sudden enlargement.

Diagnosis

- *Clinical diagnosis*—painless swelling present from birth with transillumination test positive will give clue to the diagnosis.
- *Laboratory diagnosis*—histologically, the cystic hygroma consists of dilated cystic spaces lined by endothelial cells.

Management

- *Surgical excision*—complete surgical removal of the mass.

Nasopharyngeal Cyst

They are rare clinical entities. They may be classified as congenital or acquired and in midline or lateral. They are

lined by ciliated or non-ciliated columnar epithelium with areas of squamous metaplasia in response to inflammatory stimuli. Lymphoid follicles are present in the wall.

Congenital midline cyst arises either from the pharyngeal bursa or from Rathke's pouch. Cysts arising from Rathke's pouch are exceedingly rare. They have a median base attached to the nasopharyngeal vault and lie anterior to the usual site of origin of retention and pharyngeal bursa cyst. These are lined by stratified squamous epithelium, in keeping with their ectodermal origin.

Thymic Cysts

They are rare clinical entities, which arise in persistent thymic tissue, which may occur in any location between the angle of the mandible and midline of the upper neck to the sternal notch.

Histologically, the cyst is lined by squamous and cuboidal epithelium and thymic tissues are present in the wall.

Cysts of Salivary Glands

It is described in Chapter 26: Salivary Gland Disorders.

Parasitic Cyst/Hydatid Cyst

It is also called as 'hydatid disease' or 'echinococcosis'. It is caused by the larvae *E. granulosus*, the dog tapeworm and *E. multilocularis*.

Pathogenesis

- *Ingestion of tapeworm ova*—this tapeworm lives in the intestinal tract of the dog. Its ova are excreted in the faeces of the dog and may be ingested by the intermediate host like cattle sheep and pigs. Man is also susceptible as an intermediate host as dogs are common household pets, so may accidentally ingest the ova.
- *Hatching of ova*—the ingested ova hatch in the upper gastrointestinal tract, from where the small embryo permeate the intestinal mucosa and are distributed through the blood vessels and lymphatics to all parts of the body.

Clinical Features

- *Site*—it is more common in liver, but others are found in lung, bones and brain.
- *Symptoms*—usually they are asymptomatic but they enlarge progressively to cause pressure symptoms.
- *Signs*—liver involvement causes hepatomegaly and jaundice. Pulmonary hydatid cyst may cause shortness of breath and hemoptysis.
- *Complications*—rare complication of hydatid cyst includes rupture, suppuration and calcification of the cyst.

Oral Manifestation

- **Site**—intraorally, it is found in tongue and angle of mandible.
- **Appearance**—oral hydatid cyst presents as painless, progressively increasing, soft, elastic, and well circumscribed fluctuant swelling.
- **Symptoms**—there may be speech and mastication difficulties.

Radiographic Features

- There are four stages in the radiographic evolution of the disease. They are as follows:
 - **First stage**—there is cystic bone destruction involving medullary portion of the bone. The cysts are rounded and are described as 'grape like'. The medulla is expanded and disease is sharply demarcated.
 - **Second stage**—it is of secondary infection. In this stage, osteitis is the main change and new bone is formed beneath the periosteum. Septa between the cysts become coarser and the margins become less distinct. Pathologic fracture may occur.
 - **Third stage**—the third stage is characterized by confluent abscesses which break into the soft tissue.
 - **Fourth stage**—in fourth and final stage, bone is destroyed. The whole affected portion of the bone is represented by a 'meaningless jumble of calcareous bubbles'. The patient is likely to be crippled and bedridden.

Diagnosis

- **Clinical diagnosis**—painless, elastic and well circumscribed swelling in tongue may give clue to diagnosis.
- **Radiological features**—appearance of meaningless jumble of calcareous bubbles. In initial stage, it appears like grapes.
- **Laboratory diagnosis**—the outer layer is derived from fibrous tissue and is infiltrated by chronic inflammatory cells, eosinophils, lymphocytes and giant cells. The intermediate layer is white, non-nucleated and consists numerous delicate laminations. Innermost layer is a nucleated germinal layer. The cystic fluid is usually clear, albumin free and contains so called 'hydatid sand' consisting of brood capsule and scolices.

Management

- **Surgical**—the ideal treatment for hydatid cyst is surgical.
- **Prevention**—limiting contact between dogs and humans and deworming dogs at regular intervals are useful steps towards prevention.

Cysticercosis Cellulosae

Man develops cysticercosis through the larval form, cysticercus cellulosae, of the pork tapeworm *Taenia solium*. He can act as both the intermediate and the definitive host.

Pathogenesis

- **Ingestion of worm**—the adult worm may be ingested from inadequately heated or frozen pork. Alternatively, man may ingest the cysticerci themselves from infested pork and these develop into adult worms.
- **Attachment to wall of intestine**—these lives attach to the wall of the small intestine where they fully grow and at times, reach a length of 7 mm. Gravid proglottids or eggs begin to drop off and are passed in the faeces.
- **Penetration into intestinal mucosa**—in this way, man through contaminated food or from their own dirty hands may ingest them or they may be regurgitated into the stomach. In the stomach, their covering is digested off and the larval forms are hatched. It penetrate the intestinal mucosa and are then distributed through the blood vessels and lymphatics to all the parts of the body where they develop into cysticerci.

Clinical Features

- **Signs and symptoms**—it depends upon the site and the number of the cysticerci in the body. During the stage of invasion, there are no symptoms or slight muscular pain and mild fever may be present. CNS involvement produces serious effects.

Oral Manifestations

- **Site**—there is very little report about cysticercosis of oral region. In oral cavity, most common site involved is the tongue. Other sites involved are cheek and lips.
- **Signs**—there is a firm mass and contains watery fluid and coiled white structure apparently attached to the inner aspect of cyst.
- **Symptoms**—it is a painless, well circumscribed, elastic and fluctuant swelling.

Radiological Features

- **Appearance**—when the larvae die in the soft tissue, calcification takes place and it appears on the radiograph. The shadow of calcific density is rod shaped or resembles a "drop". Some of these appear more of less rounded when the projection of the rays happen to strike the larvae in its long axis.
- **Size**—the size of the shadow is 1 mm or more in width and 1.5 cm in length.

Diagnosis

- *Clinical diagnosis*—not specific as it is very rare in oral cavity.
- *Radiological diagnosis*—calcification is seen as 'drop' like
- *Laboratory diagnosis*—it shows dense fibrous outer capsule which is derived from host tissue. It contains a fairly dense inflammatory cell infiltrate consisting predominantly of lymphocytes, plasma cells and histiocytes. Few foci of dystrophic calcification are present in this capsule and some of these are concentrically laminated. The cyst lies within this membrane and contains larval form of *T. solium*.

Management

- *Surgical*—cutaneous cyst should be surgically removed.
- *Mebendazole*—Mebendazole has some value in the treatment.
- *Prevention*—pork should be properly cooked as preventive measures.

Syndromes Associated with Odontogenic Cysts

Jaw Cyst-Basal Cell Nevus-Bifid Rib Syndrome

It is also called as 'nevoid basal cell carcinoma', 'Gorlin's and Goltz's syndrome'. It is transmitted as autosomal dominant trait with poor degree of penetration and variable expressivity. It is caused by mutation in patched (PTCH), tumor suppressor gene.

Clinical Features

- *Age*—it appears early in life after 5 years and before 30 years.
- *Basal cell carcinoma*—Nevoid basal cell carcinoma is brownish colored papules predominant on skin, neck and trunk. Basal cell carcinoma is less aggressive in this syndrome than in solitary basal cell carcinoma.
- *Cutaneous abnormalities*—dermal cyst and tumors, palmar pitting, palmar and plantar keratosis and dermal calcinosis. Skin lesions are small, flattened, flesh colored or brownish papules occurring anywhere in the body, but are prominent on the face and trunk.
- *Skeletal abnormalities*—rib anomalies and brachymetacarpalism, bifid rib, agenesis, deformity and synostosis of rib; kyphoscoliosis, vertebral fusion, polydactyly and shortening of metacarpals.
- *Skull features*—there is frontal and temporoparietal bossing, resulting in an increased cranial circumference.
- *Ophthalmologic abnormalities*—hypertelorism with wide nasal bridge, dystopia canthorum, congenital blindness and internal strabismus.

- *Neurological anomalies*—mental retardation, calcification of falx cerebri and other parts of dura, agenesis of corpus callosum, congenital hydrocephalus and occurrence of medulloblastomas.
- *Sexual abnormalities*—Hypogonadism in males and ovarian tumors.

Oral Manifestations

- *Odontogenic keratocyst*—jaw lesions appear as multiple odontogenic keratocysts usually appearing in multiple quadrants.
- *Mandibular prognathism*—mild mandibular prognathism is also present in this syndrome.

Radiographic Features

- *Odontogenic keratocyst*—jaw cysts appear as a multiple cysts like radiolucency of variable size varying from few millimeters to several centimeters (**Fig. 13-47**). They occur most frequently in premolar-molar region.
- *Calcified falx cerebri*—radiopaque lines of calcified falx cerebri are prominent on PA projection.

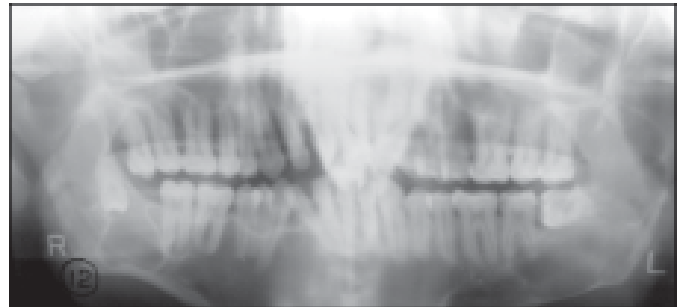


Fig. 13-47: Multiple OKC seen in Gorlin's syndrome (Courtesy Dr Ashok L).

Differential Diagnosis

- *Multiple myeloma*—'Bence Jones' protein in the urine.
- *Metastatic carcinoma*—history of primary tumor.
- *Histiocytosis X*—does not have characteristic borders as seen in cyst.
- *Cherubism*—jaw expansion is common in it.

Management

- *Enucleation*—complete enucleation of the lesion should be done. Periodic examination should be done to check for recurrence.

Treatment of Cysts

Regression of Cysts without Surgical Treatment

There is good evidence that some small radicular cyst will regress if the necrotic pulp remnant and bacteria are

removed from the root canal of the causative tooth and the canal is effectively filled.

Circumscribed rounded zone of periapical bone destruction perhaps up to 1.5 cm in diameter will be seen in some patients to reduce in size and resolve over a period of month.

Marsupialization of Dental Cysts

It is known as '*Partsch's operation*' as it is discovered by Partsch. It involves making an opening into the cyst as large as practical and packing the cavity. It is a more effective way of emptying a cyst.

Indication

- *Age*—in a young child, with developing tooth germs.
- *Proximity to vital structure*—when proximity of the cyst to vital structure can create an oronasal or oroantral fistula or injure the neurovascular structure, this treatment modality should be considered.
- *Eruption of teeth*—in a young patient with a dentigerous cyst, marsupialization will permit the eruption of the unerupted tooth or any other developing tooth which has been displaced.
- *Vitality of teeth*—when the apices of many adjacent erupted teeth are involved within a large cyst, enucleation can prejudice the vitality of these teeth.
- *Fracture in cystic region*—it is the simplest way to treat a fracture complicating a large cyst of the mandible.

Advantages

- *Simple procedure*—this procedure at least for large cysts, is technically simple.
- *No general anesthesia required*—even quite large cysts can be dealt with under local anesthesia. As anesthesia of deeper recesses is not essential and this is particularly an advantage in the maxilla.
- *Preservation of adjacent mucosa*—because the deeper part of the lining is not disturbed, adjacent important structures are not put at the risk, i.e. the blood vessels to the apices of adjacent vital teeth, the inferior dental neurovascular bundle and the integrity of lining of the antrum or nose are well preserved.
- *Conservation of tooth*—marsupialization may be the best way to conserve the tooth of origin of a dentigerous cyst and to permit its eruption.
- *Prevention of fistula*—it prevents oronasal and oroantral fistula.
- *Reduce operating time and blood loss*—it reduces operating time and blood loss and helps in the shrinkage of cystic lining.
- *Preservation of alveolar ridge*—alveolar ridge is preserved.

Disadvantages

- *Regular postoperative care*—the need for regular postoperative care, possibly over a substantial period of time. It is necessary to supervise healing so that opening remains large in proportion to the underlying cavity. Prolonged follow-up visits and periodic irrigation.
- *Problems of opening*—a rapid reduction in the size of the opening may be difficult to prevent. Indeed an opening into a large cyst in the ramus may present significant problems.
- *Uneven reduction*—uneven reduction in size of the cavity may result in slit-like cavity, difficult to keep clean.
- *New cyst formation*—there is risk of invagination and new cyst formation.
- *Adjustment of plug*—regular adjustments of plug.

Enucleation

Enucleation allows for the cystic cavity to be covered by a mucoperiosteal flap and the space fills with blood clot which will eventually organized and form normal bone.

Indications

- *Odontogenic keratocyst*—it is indicated in treatment of odontogenic keratocyst.
- *Recurrent lesion*—recurrence of cystic lesion of any cyst type.

Advantages

- *Primary closure*—as the complete cyst is removed, primary closure of the wound is possible.
- *Rapid healing*—healing is rapid due to primary closure.
- *Less postoperative care*—postoperative care is reduced.
- *Thorough examination*—thorough examination of the entire cystic cavity can be done.

Disadvantages

- *Healing cannot be observed*—after primary closure, it is not possible directly to observe the healing of the cavity as with marsupialization.
- *Removal of unerupted teeth*—in young persons, the unerupted teeth in a dentigerous cyst will be removed with the lesion in this mode of treatment.
- *Mandibular weakening*—removal of large cysts will weaken the mandible making it prone to jaw fracture.
- *Damage of adjacent structure*—it will damage to adjacent vital structures.

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14

Odontogenic Tumor of Jaw

Classification

First Classification

Benign

Epithelial

- With inductive changes in the connective tissue
 - Ameloblastofibroma
 - Dentinoma
 - Calcifying odontogenic tumor
 - Odontoameloblastoma
 - Odontoma
- Without inductive changes in the connective tissue
 - Ameloblastoma
 - CEOT
 - Epithelial atypia
 - Ameloblastic changes in odontogenic cyst

Mesenchymal

- Odontogenic myxoma
- Odontogenic fibroma
- Cementoma
- Periapical cemental dysplasia
- Cementifying fibroma
- Benign cementoblastoma

Malignant

Epithelial

- With inductive changes in the connective tissue
 - Ameloblastic fibrosarcoma
 - Ameloblastic odontosarcoma
- Without inductive changes in the connective tissue
 - Malignant ameloblastoma
 - Primary intraosseous carcinoma
 - Malignant changes in odontogenic cyst

Second Classification (By WHO)

Benign

Odontogenic epithelium without odontogenic ectomesenchyme

- Ameloblastoma
- Squamous odontogenic tumor
- Pindborg's tumor
- Clear cell odontogenic tumor

Odontogenic epithelium with odontogenic ectomesenchyme with or without dental hard tissue formation

- Ameloblastic fibroma
- Ameloblastic fibro-odontoma
- Ameloblastic fibro-dentinoma
- Odontoameloblastoma
- Adenomatoid odontogenic tumor
- Complex and compound odontoma

Odontogenic ectomesenchyme with or without including odontogenic epithelium

- Odontogenic fibroma
- Odontogenic myxoma
- Benign cementoblastoma

Malignant tumor

Odontogenic carcinoma

- Malignant ameloblastoma
- Primary intraosseous carcinoma
- Malignant variant of other odontogenic epithelial tumors
- Malignant changes in odontogenic cyst

Odontogenic sarcoma

- Ameloblastic fibrosarcoma
- Ameloblastic fibrodentinosa sarcoma

Third Classification (Benign Tumor)

Epithelial

- Ameloblastoma
- Adenoameloblastoma
- Enameloma
- Pindborg's tumor (CEOC)

Mesenchymal

- Dentinoma
- Cementoma
- Cementoblastoma
- Odontogenic fibroma

Mixed

- Ameloblastic fibroma
- Ameloblastic fibroodontoma
- Ameloblastic odontoma
- Odontogenic myxoma
- Compound composite odontoma
- Complex composite odontoma
- Odontogenic fibroma

Developmental

- Dense invaginatus or dilated odontome
- Dense evaginatus

Fourth Classification

Lesions consisting of odontogenic epithelium

- Ameloblastoma
- Adenomatoid odontogenic tumor
- Calcifying epithelial odontogenic tumor
- Calcifying odontogenic cyst

Lesions consisting of odontogenic epithelium and mesenchyme

- Ameloblastic fibroma
- Ameloblastic sarcoma

Lesions consisting of odontogenic epithelium and calcified dental tissues

- Odontoameloblastoma

Lesions consisting of calcified dental tissue, but without odontogenic epithelium

- Complex odontome
- Compound odontome
- Dens invaginatus (dilated odontome)
- Enameloma
- Dentinoma
- Cementoma

Lesions consisting of odontogenic mesenchyme

- Odontogenic fibroma
- Odontogenic myxoma

Benign Tumors

Ameloblastic Fibroma

It is also called as '*fibrous adamantinoma*', '*soft odontoma*', '*soft mixed odontoma*', and '*fibroadamantoblastoma*'. It is characterized by simultaneous proliferation of both epithelial and mesenchymal tissue without formation of enamel and dentin. If this tumor is left undisturbed, it will ultimately differentiate into a lesion known as '*ameloblastic fibroodontoma*'. It may mature into a complex odontoma. It is characterized by neoplastic proliferation of maturing and early functional ameloblasts as well as the primitive mesenchymal components of the dental papilla.

Origin

- *Dental follicle*—it develops from the dental follicle usually after the onset of calcification of the tooth.
- *Tooth bud*—in some instances, it may develop from tooth bud before onset of calcification and abort the formation of normal tooth.

Clinical Features

- *Age and sex distribution*—average age of occurrence is under 20 years with 40% of patients under the age of 10 years. There is slight predilection for occurrence in males.
- *Site*—it is developed in premolar, molar area of the mandible (**Fig. 14-1**).
- *Symptoms*—it is painless and expands slowly. There is bulging of the cortical plates rather than erosion through them. There is also migration of involved teeth.
- *Signs*—it enlarges by gradual expansion so that the periphery of bone often remains smooth. It is associated



Fig. 14-1: Swelling seen in premolar area in mandible in ameloblastic fibroma (Courtesy Dr Parate).

with unerupted teeth. It has got a slower clinical growth than ameloblastoma and does not tend to infiltrate between trabeculae of bone.

- **Size**—it reaches upto an approximate size of 1 to 8.5 cm.

Radiographic Features

- **Site**—it develops in premolar-molar area of the mandible. In some cases, it may involve the ramus and extend forward to the premolar-molar area.
- **Radiodensity**—it may present with cyst-like area of bone destruction or there may wide area of bone destruction (**Fig. 14-2**). As it is usually associated with an impacted tooth, it appears as pericoronal radiolucency.
- **Internal structure**—it may be either unilocular or multilocular and is associated with unerupted or missing tooth.
- **Margins**—are well defined often with sclerotic borders.
- **Effect on surrounding structure**—buccal and lingual expansion of jaw with intact cortical plates. Teeth may be inhibited from normal eruption or may be displaced in an apical direction.

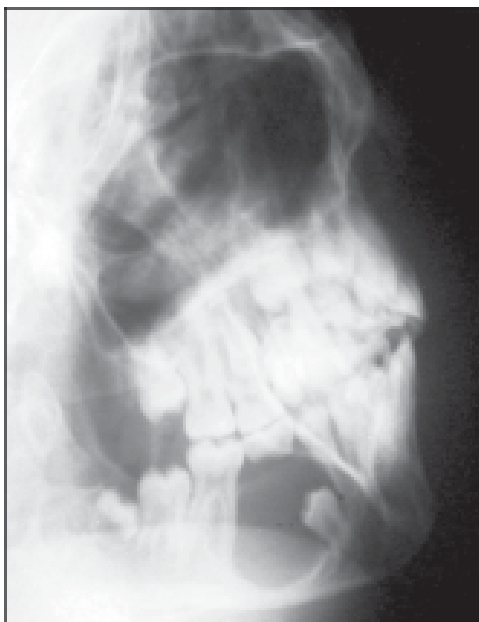


Fig. 14-2: Cyst-like radiolucency seen in case of ameloblastic fibroma (Courtesy Dr Datarkar).

Diagnosis

- **Clinical diagnosis**—not so specific.
- **Radiological diagnosis**—not so specific. Sometimes, CT scan may be helpful to diagnose the tumor (**Fig. 14-3**).
- **Laboratory diagnosis**—biopsy shows scattered islands of epithelial cell in a rosette, long finger-like strands, nests and cords-like pattern. Cells resemble the primitive odontogenic epithelium. The mesenchymal component is made up of a primitive connective tissue.



Fig. 14-3: CT scan of ameloblastic fibroma (Courtesy Dr Bhaskar Patle).

Differential Diagnosis

- **Central giant cell granuloma**—it has a honeycomb pattern and is usually seen in anterior location.
- **Odontogenic myxoma**—finer trabeculation resembling a string of tennis racket pattern.
- **Central hemangioma**—honeycomb pattern and local gingival bleeding and pumping action of tooth or teeth.
- **Primordial cyst**—thick yellowish granular fluid.
- **Ameloblastoma**—associated with much older group.

Management

- **Curettage**—it can be done which has successful results in many cases. But there are chances of recurrence in this tumor.
- **Surgical excision**—aggressive surgical excision should be done for recurrent lesion.

Calcifying Epithelial Odontogenic Tumor

It is also called as '*Pindborg's tumor*' or '*calcifying ameloblastoma*'. It arises from the reduced enamel epithelium or dental epithelium. They are located within the bone and produce mineralized substance like amyloid. It forms 1% of all odontogenic tumors.

Clinical Features

- **Age and sex**—it is more common in men with an age range of 8 to 92 years with a mean age of 42 years.
- **Site**—mandible is more commonly affected than the maxilla in the ratio of 2:1 and developed in premolar-molar area (**Fig. 14-4**).



Fig. 14-4: Swelling seen in mandibular region due to calcifying epithelial odontogenic tumor (Courtesy Dr Bhaskar Patle).

- **Symptoms**—it is asymptomatic and only presenting symptom is a painless swelling. In rare cases, there is associated mild paresthesia.
- **Signs**—cortical expansion occurs. Palpation will show hard tumor with well defined or diffuse border. It is locally invasive with a high recurrence rate.
- **Peripheral calcifying epithelial odontogenic tumor**—there are only few reported cases of this type. It is presented as sessile gingival growth, seen on anterior gingiva (Fig. 14-5).



Fig. 14-5: Calcifying epithelial odontogenic tumor showing swelling in premolar area (Courtesy Abhishek Soni).

Radiographic Features

- **Radiodensity**—it may be totally radiolucent to mostly radiopaque area around the crown of unerupted teeth. The radiopacity is produced by mineralization of amorphous proteinaceous material generated by the tumor cells rather than the disorganized formation of dental tissue (Fig. 14-6).

- **Margin**—it may or may not be well demarcated from the surrounding normal tissue. It has got scalloped margin.
- **Internal structure**—there may be a combined pattern of radiolucency and radiopacities with many small, irregular bony trabeculae traversing the radiolucent area in many directions producing a multilocular or honey-comb pattern.

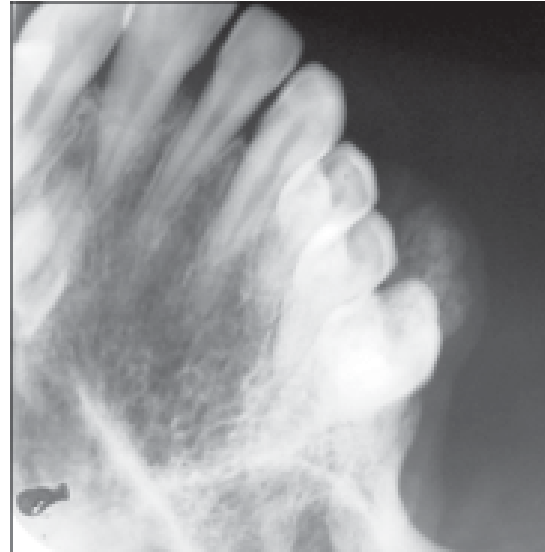


Fig. 14-6: Radiopaque calcified structure seen in CEOT (Courtesy Dr Tapasya Karamore).

- **Driven snow appearance**—later, it reveals a unilocular or a multilocular cystic lesion with numerous scattered radiopaque foci of varying sizes and density which gives it an appearance as that of 'driven snow' (Fig. 14-7).

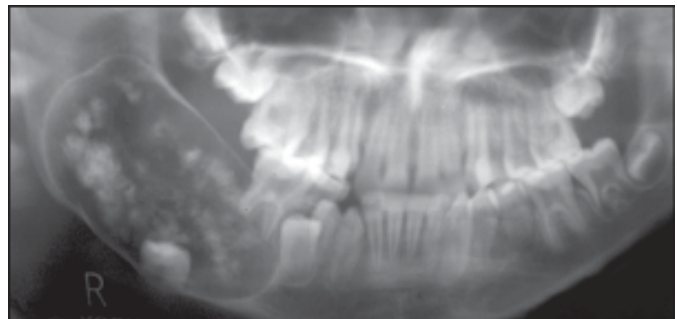


Fig. 14-7: Typical driven snow appearance seen in CEOT (Courtesy Dr RN Mody).

- **Effect on surrounding structure**—it may displace the developing tooth or prevent its eruption. Expanded cortical plate can be visualized in buccal, lingual and vertical dimension.

Diagnosis

- **Clinical diagnosis**—not specific.
- **Radiological diagnosis**—typical driven snow appearance is seen.

- *Laboratory diagnosis*—biopsy shows polyhedral epithelial cells packed in large sheets. There is presence of calcification in large amounts and often in the form of 'Liesegang rings'.

Differential Diagnosis

- *Central odontogenic fibroma*—Histopathologically, fibroblasts are prominent and abundant.
- *Calcifying epithelial odontogenic cyst*—aspiration yield viscous granular yellow fluid.
- *Adenomatoid odontogenic tumor*—it is more common in anterior maxilla as compared to calcifying epithelial odontogenic tumor which is common in mandibular premolar-molar area.
- *Partially calcified odontoma*—it appears within the capsule.

Management

- *Conservative surgical excision*—it has limited invasive potential therefore local excision with limited margins is indicated. Simple enucleation can be done.

Odontoma

It is a hamartoma of odontogenic origin in which both epithelial and mesenchymal cells exhibit complete differentiation with enamel, dentin and cementum that are laid down in abnormal position. Odontome may arise from any of three dental tissues, i.e. enamel, dentin and cementum.

Origin

- There is growth of epithelial and mesenchymal component exhibiting complete differentiation with a result that functional ameloblasts and odontoblasts laid down in abnormal fashion.

Etiology

- *Genetic transmission*—it may occur due to mutant gene or interference with genetic control of tooth development.
- *Local trauma or infection*—in some cases of trauma or infection to tooth in developing stage may cause some alteration in ectomesenchymal interaction causing odontoma formation.

Classification

Ectodermal origin

- *Enameloma* (enamel pearl, enamel nodule).

Mesodermal origin

- *Dentinoma*
- *Cementoma*.

Mixed

- *Complex composite odontoma*—non-discrete masses of dental tissue.
- *Compound composite odontoma*—multiple well formed teeth.
- *Compound-complex odontoma*—some tumors contain not only multiple teeth like structures, but also calcified masses of dental tissue in a haphazard manner, such lesion are called as compound-complex odontoma.
- *Geminated odontoma*.
- *Dilated odontoma with dens in dente*.

Enameloma

It is also called as 'enamel nodule', 'enamel drop' or 'enamel pearl'. It is a small focal excessive mass of enamel on the surface of the tooth. It is formed by small group of misplaced ameloblasts. This enamel may contain a small core of dentin and rarely, small strands of pulp tissue extending from pulp chamber or root canal of the tooth.

Clinical Features

- *Site*—enamel pearls are most commonly found in the bifurcation or trifurcation of roots or on the surface near the cemento-enamel junction. The most common site is upper molar.
- *Appearance*—it appears as a tiny globule of enamel firmly adherent to the tooth.
- *Significance*—in some cases, it occupies such a position on cemento-enamel junction that leads to pocket formation which may predispose to periodontal diseases.

Radiographic Features

- *Radiodensity*—the little mass is made up of enamel which absorbs more X-rays than the adjacent dentin so that it appears as radiopaque shadow.
- *Shape*—it is usually hemispherical.
- *Size*—it varies in size from one millimeter to three or more.
- *Appearance*—it appears as rounded, well circumscribed density associated with root.
- *Dense projection*—it is best seen radiographically when it is situated on mesial and distal side of the tooth so that it appears in profile. It then appears as dense projection which is continuous with the tooth substance.

Diagnosis

- *Clinical diagnosis*—tiny enamel globule seen on enamel.
- *Radiological diagnosis*—radiopaque shadow on enamel.
- *Laboratory diagnosis*—it consists of enamel.

Differential Diagnosis

- *Projection shadow*—it happens when the angulation of the rays is such that they do not pass freely between the roots where they approximate at the bifurcation. The result is that portion of distal aspect of the mesial root is projected over the mesial portion of distal root. It will add radiographic density which results in more or less rounded shadow of greater density where the roots converge at the bifurcation. To differentiate it from enameloma, take radiograph in such a way that the rays should pass between the contiguous portions of the roots.

Management

- *Removal of mass*—mass has to be removed if it is causing periodontal diseases.

Dentinoma

It is a rare tumor of odontogenic origin composed of immature connective tissue, odontogenic epithelium and irregular or dysplastic dentin.

Clinical Features

- *Age and sex*—it is seen in patients younger than 36 years with an average age of 26 years with no sex predilection for occurrences.
- *Site*—it is predominately seen in the mandibular molar area and is frequently associated with an impacted tooth.
- *Symptoms*—patient notices a swelling over a variable period of time with pain.
- *Sign*—perforation of mucosa and subsequent infection may be present. There may be redness of overlying mucosa with discharge.

Radiographic Features

- *Radiodensity*—radiopaque mass associated with the crown of unerupted tooth.
- *Internal structure*—it contains either a large, solitary, opaque mass or numerous smaller, irregular radiopaque masses of calcified materials which may vary considerably in size.
- *Bone*—it may cause local destruction of bone.

Diagnosis

- *Clinical diagnosis*—not specific.
- *Radiological diagnosis*—radiopaque mass with crown of tooth.
- *Laboratory diagnosis*—biopsy shows irregular dentin, which has been termed as dentinoid or osteodentin. There is also presence of undifferentiated odontogenic epithelium.

Management

- *Surgical*—surgical excision with thorough curettage of the area.

Complex and Compound Odontoma

It is a non-aggressive lesion that is more likely to be a hamartoma than a neoplastic growth.

Etiology

- *Trauma*—local trauma or infection may lead to production of such a lesion.
- *Genetic*—they are either inherited or are due to a mutant gene or interference, possibly postnatal, with genetic control of tooth development.

Mechanism

- *Differentiation of epithelial and mesenchymal cells*—both the epithelial and mesenchymal cells exhibit complete differentiation with the result that functional ameloblasts and odontoblasts form enamel and dentin.
- *Pattern*—it is laid down in an abnormal pattern because of failure of cells to reach the morphodifferentiation stage. Lesion is composed of more than one type of tissue, for this reason it is called as composite odontome.
- *Compound composite odontome*—in some composite odontomes, the enamel and dentin are laid down in such a fashion that the structure bears a considerable anatomical resemblance to that of normal teeth except they are often smaller than the typical teeth, which have been termed as compound composite odontome.
- *Complex composite odontome*—when calcified dental tissues are simply arranged in an irregular mass bearing no morphological similarity even to rudimentary tooth then that form is called as complex composite odontome.

Clinical Features

- *Age and sex*—it is more commonly seen in first and second decade of life. Most begin to form while normal dentition is developing. Slight predilection for occurrence in males.
- *Site*—compound odontome occurs in incisor, canine area of maxilla and complex odontome occurs in mandibular 1st and 2nd molar area. Unusual situation includes the maxillary sinus, inferior border of the mandible, ramus and condylar region.
- *Frequency*—compound odontome is twice as common as complex odontome.
- *Size*—compound odontoma is between 1 to 3 cm in diameter. It usually remains small and diameter of the mass only occasionally increases than that of the tooth.

- **Symptoms**—patient may notice alveolar swelling in the jaw. This swelling will cause facial asymmetry (**Fig. 14-8**). In some cases, signs of infection may be present.



Fig. 14-8: Odontoma causing swelling on right side of face causing facial asymmetry (Courtesy Dr Parate).

- **Signs**—it is common for a tooth or teeth to be absent from the arch in the presence of an odontome. On palpation expansion of the jaw (**Fig. 14-9**) may be noticed.



Fig. 14-9: Intraoral picture of same patient (**Fig. 14-8**) showing expansion of alveolar bone (Courtesy Dr Parate).

- **Teeth**—impaction malpositioning, diastema, aplasia, malformation and deviation of adjacent teeth are associated with 70% of odontoma.
- **Development of cyst**—sometime, cyst develops in relation with a complex odontome and compound odontome, but it is very rare.

Radiographic Features

General

- **Site**—it is situated between the roots of teeth.

- **Appearance**—it appears as an irregular mass of calcified material surrounded by narrow radiolucent bands with a small outer periphery.

Compound composite odontome

- **Appearance**—compound type shows number of teeth like structures in the region of the canine (**Fig. 14-10**).



Fig. 14-10: Compound odontoma showing shapeless tooth structure (Courtesy Dr Parate).

- **Internal structure**—there is cluster of small shapeless dense masses of solid tissue having equal or more density, depending on the size of the mass. There may be many masses each of which has own dark line surrounding it. In some cases, there may be presence of two or more teeth-like masses having conical enamel-capped crowns and with fusion of the radicular portion.
- **Characteristic of teeth**—if only few teeth are present, a periodontal and pericoronal characteristic of unerupted teeth may be discernible on each individual tooth. If a large number of teeth are present, the radiopaque mass is surrounded by a radiolucent line that represents the pericoronal space of the unerupted teeth.
- **Margin**—borders are well defined in both the cases but vary from smooth to irregular and may have hyperostotic borders.

Complex composite odontome

- **Appearance**—complex composite odontoma appears as a dense radiopaque object sometimes lying in clear space (**Fig. 14-11**). Density is greater than that of bone and to greater than or equal to the teeth.
- **Internal structure**—it shows a well defined radiolucency containing irregular masses of calcified tissue. It is associated with unerupted teeth (**Fig. 14-12**).

Cystic odontome

- **Cystic complex odontome**—it will reveal the solid mass of the odontome but without any associated unerupted



Fig. 14-11: Complex odontome showing well defined radiopacity (Courtesy Dr Parate).



Fig. 14-12: Complex odontome showing radiopacity but not resembling tooth structure.

tooth. There is dark shadow which represents the cystic cavity and it has well defined and corticated margins except when infection supervenes (**Fig. 14-13**).

- *Cystic compound odontome*—there is an area of bone destruction which appears as a dark shadow having well defined margins lined by a thin white layer of cortical border. Within the cystic cavity there are numerous white opacities which vary in size and shape. In some cases, there are small amorphous granular densities scattered throughout the cavity and in some cases there are small denticles with enamel caps or without any distinction of the component mass.

Differential Diagnosis

- *Difference between compound and complex type*—compound odontome has more than one fragment while in complex, it is one solid mass. Compound is seen most commonly

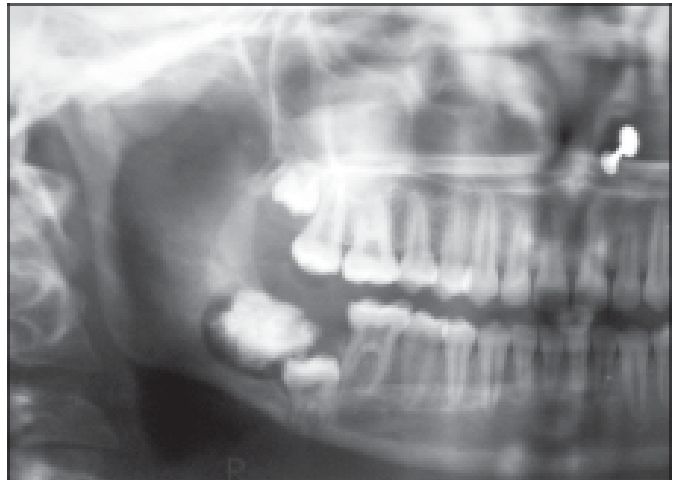


Fig. 14-13: Cystic complex odontome showing radiopacity surrounded by radiolucent capsule (Courtesy Dr Bhaskar Patle).

in maxillary canine and incisor region while complex is more commonly seen in mandibular molar area.

- *Cementifying or ossifying fibroma*—odontoma is associated with unerupted molar teeth and more radiopaque than fibroma and discovered at much younger age than fibroma.
- *Adenomatoid odontogenic tumor*—rarely as opaque as the complex type and found in association with maxillary canines.
- *Periapical cemental dysplasia*—smaller than complex type and is limited to mandibular anterior region.
- *Calcifying epithelial odontogenic tumor*—rare, less opaque and develops in the midline.
- *Fibrous dysplasia*—mottled or smoky pattern poorly defined borders.

Diagnosis

- *Clinical diagnosis*—not specific.
- *Radiological diagnosis*—compound shows shapeless mass of tooth structure while complex show disorganized mass of tooth structure.
- *Laboratory diagnosis*—biopsy shows multiple tooth structures contain in loose fibrous matrix (compound) and mature tubular dentin (complex).

Management

- *Local excision*—simple local surgical excision is the treatment of choice.

Gemination and Geminated Composite Odontome

These two terms appear to be used interchangeably with each other. The attempt at division of a single tooth germ by an invagination, with resultant incomplete formation of two teeth is gemination (**Fig. 14-14**), while the addition of

more or less misshapen one or both components may be termed as geminated composite odontome. Geminatio is discussed in detail in Chapter 9: Teeth Anomalies.



Fig. 14-14: Invaginated odontome (Courtesy Dr Mody).

Ameloblastoma

First detailed description of ameloblastoma was given by 'Falkson' in 1879. It is an aggressive tumor that appears to be arising from remnants of dental lamina or dental organs. It represents 1% of all oral tumors and 11% of odontogenic tumors. It is the most common epithelial tumor producing minimal inductive changes.

There are different names given to this tumor. Term 'adamantine epithelioma' was given by Malassez in 1885. It was replaced by ameloblastoma in 1934 by Churchill. It is also called as 'adamantinoma', 'adamantinoblastoma', 'epithelial odontome' and 'multilocular cyst'.

Definition

It is a benign but locally invasive polymorphic neoplasm consisting of proliferating odontogenic epithelium which is usually in a follicular or plexiform pattern, lying in fibrous stroma.

Etiology

- **Irritation**—it might be considered as one of the etiological factors as it often occurs in the posterior region of the mandible which is most susceptible to irritation.
- **Infection**—Robinson found that 1/3rd of the cases have history of oral infection, extraction of teeth and injuries to teeth.

- **Trauma**—trauma can be a causative factor for ameloblastoma.
- **Dietary deficiency**—dietary deficiency has been considered to be a possible factor, e.g. pronounced defect in development of tooth germ as seen in rickets may lead to irregularity in the ameloblastic layer.
- **Virus**—injection of polyoma viruses and nitrosoureas have been shown to produce ameloblastoma-like lesion in animals.

Classification

- **On pathological basis**
 - Conventional ameloblastoma
 - Unicystic ameloblastoma
 - Peripheral ameloblastoma
 - Pituitary ameloblastoma
 - Adamantinoma of long bones
- **On histological type**
 - Follicular ameloblastoma
 - Plexiform ameloblastoma
 - Acanthomatous ameloblastoma
 - Basal cell ameloblastoma
 - Unicystic ameloblastoma
 - Plexiform unicystic ameloblastoma
 - Granular cell ameloblastoma
 - Papilliferous ameloblastoma
 - Hemangioameloblastoma
 - Desmoplastic ameloblastoma
 - Clear cell ameloblastoma
 - Dentinoameloblastoma
 - Melanoameloblastoma
 - Keratoameloblastoma.

Pathogenesis

The resemblance of the tumor epithelium to the normal enamel organ indicates that ameloblastoma arises from dental epithelium. The possibilities for its development are as follows.

- **Enamel organ**—due to histological similarities such as its origin, it has been thought that the tumor growth starts at an early age; i.e. during the period of existence of enamel organ but most of the patients are middle aged i.e. in a period which is long after regression of the enamel organ. However, the occasional occurrence of the tumor as a unilocular cystic lesion surrounding the crown of an unerupted tooth also suggests that in some cases, the enamel organ may give rise to it.
- **Cell rests**—cell rests of enamel organ either remnants of dental lamina or remnants of Hertwig's sheath i.e. epithelial cell rests of Malassez have the potential of transforming into ameloblastoma.

- *Epithelium of odontogenic cysts, i.e. from dentigerous cyst and odontoma*—this may be possible as the epithelium in the wall of the cyst and that of ameloblastoma are derived from the same embryonic source.
- *Oral mucosa*—basal cells layer of the oral epithelium of jaws can be the origin. The reason behind this is that there is communication between the tumors and overlying mucosal epithelium as seen in peripheral ameloblastoma.
- *Heterotrophic epithelium*—heterotrophic epithelium in other parts of the body especially of pituitary gland can serve as source of origin.

Clinical Features

- *Incidence*—ameloblastoma accounts for approximately 1% of all oral tumors and 11% of all odontogenic tumors.
- *Sex and race*—it has slight predilection for males and often seen in blacks.
- *Age*—most patients are between 20 to 50 years of age with mean age of discovery being 40 years. The tumor can occur in young children. Unicystic type of ameloblastoma is more common in the 2nd and 3rd decade and the extrasosseous form is more common in the older age group.
- *Site*—it develops in the molar ramus area (approximately $\frac{3}{4}$ th of cases) in the mandible and also occurs in maxilla in third molar area, followed by the maxillary sinus and floor of the nose. The right side of the mandible is affected slightly more as compared to the left side.
- *Preceding factors*—neoplasm is frequently preceded by extraction of teeth, cystectomy and some other traumatic episodes.
- *Onset*—it begins as a central lesion of the bone which is slowly destructive but tends to expand bone rather than perforating it.
- *Symptoms*—patient notices a gradually increasing facial asymmetry (**Fig. 14-15**). Teeth in involved region are displaced and become mobile (**Fig. 14-17**). Pain and paresthesia may occur, if the lesion is pressing upon a nerve or is secondarily infected.
- *Signs*—in later stages, the lesion may show ovoid and fusiform enlargement that is hard but non-tender. If it is left untreated for many years the expansion may be extremely disfiguring (**Fig. 14-16**), fungating and ulcerative type of growth characteristic of carcinoma can be seen.
- *Eggshell cracking*—as tumor enlarges, palpation may elicit a hard sensation or crepitus. Surrounding bone may become thin so that fluctuation and 'eggshell cracking' may be elicited.
- *Maxillary ameloblastoma*—maxillary lesions are more dangerous than mandibular lesions due to tendency for



Fig. 14-15: Facial asymmetry in patient of ameloblastoma.



Fig. 14-16: Disfigurement of face due to huge swelling of ameloblastoma (Courtesy Dr Bhaskar Patle).



Fig. 14-17: Intraoral view of ameloblastoma showing displacement of tooth.

the former lesion to spread more extensively in the more porous maxillary bone and possibility of the involvement of the cranial base. It may extend into the paranasal air sinuses, orbit or the nasopharynx.

- **Mural ameloblastoma**—ameloblastoma may form from the epithelial lining of dentigerous cyst and in such cases, it is called as '*mural ameloblastoma*'.
- **Spread**—this tumor causes expansion of the bone rather than destruction. Ameloblastoma infiltrates cancellous bone but it is largely confined by the compact bone. It never infiltrates haversian canal. It infiltrates between the trabeculae of the cancellous bone and hence leaves them intact for some duration. Hence, the actual margins of the lesion are much greater than radiological margin which accounts for recurrence rate, if neglected. As the cortical plates of maxilla are thin, it does not possess an effective barrier to the spread of ameloblastoma into adjacent vital structures.

Radiographic Features

- **Radiodensity**—in early stages, there is area of bone destruction which is well defined and is indicative of slow growth with hyperostotic borders.
- **Margins**—outline is smooth, scalloped, well defined and well corticated. The walls of the cavity are coarse. In some cases the margins of the tumor are lobulated.
- **Internal structure**—usually it is multilocular but may be unilocular (**Fig. 14-18**). Coarse or fine trabeculae may be present within the tumor and it is common for the free margins of the tumor to be devoid of bony covering.



Fig. 14-18: Unilocular ameloblastoma seen in posterior region of mandible.

- **Appearance**—there is presence of septa in the lesion. In some cases, number and arrangement of septa may give

the area a '*honeycomb appearance*' (**Fig. 14-20**) (numerous small compartments) or a '*soap bubble appearance*' (larger compartments) (**Fig. 14-19**). In advanced stages, perforated cortical plate may contribute to a multilocular appearance.

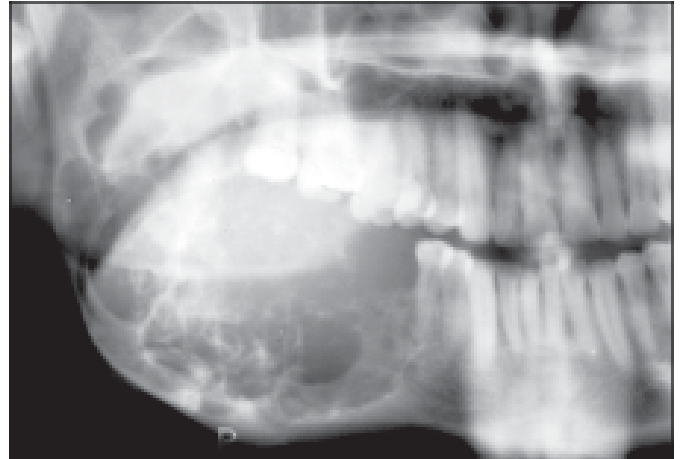


Fig. 14-19: Soap bubble appearance seen in ameloblastoma.

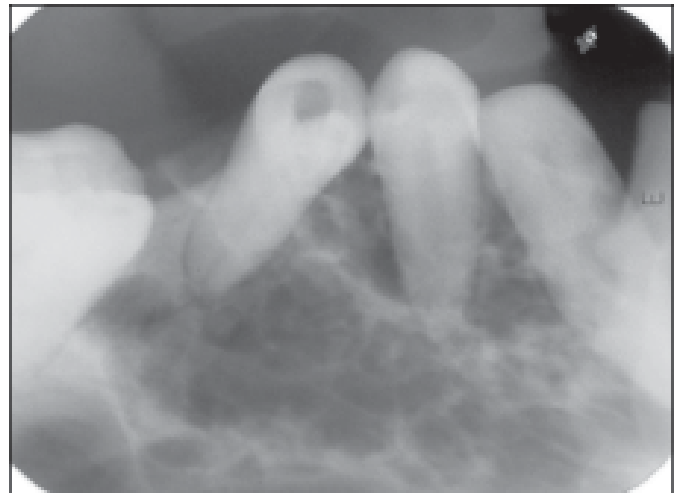


Fig. 14-20: Honeycomb appearance seen in ameloblastoma.

- **Progress**—in early stage ameloblastoma presents as bubble like appearance with fairly large, round and distinct compartments. As the tumor grows and expands, the compartments may coalesce and fuse. As the lesion again increases in size, the cortex is expanded and destroyed.
- **Subclinical lesion**—a small subclinical lesion usually presents in radiographs as many small rounded cavities in the bone having sharply defined and sometimes corticated borders. In some cases, there are two rounded and well defined small cavities having good bony cortex; in the center of which there is a small white dot.
- **Unicystic variety**—in case of mural ameloblastoma, the radiological appearance is like unicystic variety (**Fig. 14-21**).

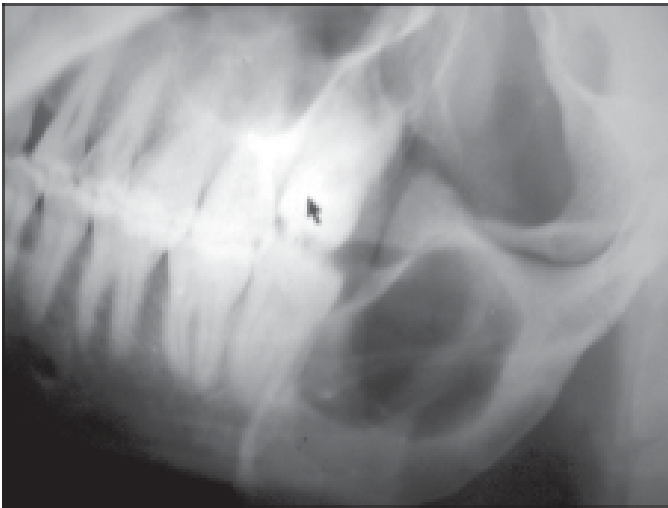
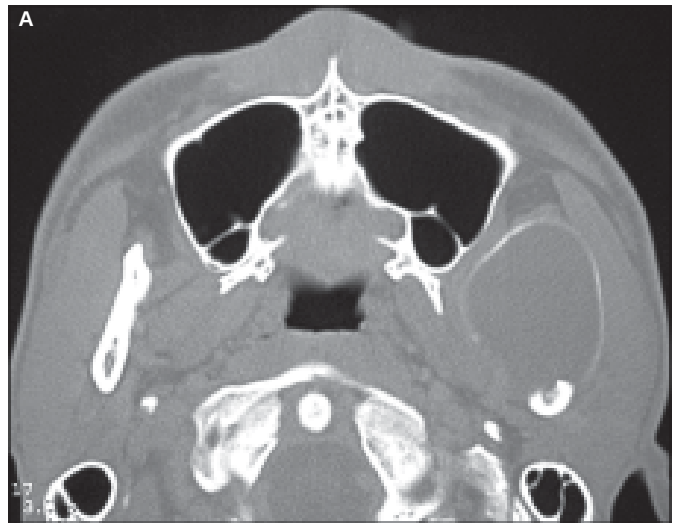


Fig. 14-21: Unilocystic ameloblastoma (Courtesy Dr Mody).



Fig. 14-22: Ameloblastoma also showing soft tissue swelling in the radiograph (Courtesy Dr Bhaskar Patle).



Figs 14-23A and B: CT scan of ameloblastoma (Courtesy Dr Iswar).

- *Effect on surrounding structures*—the jaws are likely to be enlarged, depending on the overall size of the tumor. Extensive root resorption may occur. Thickening of membrane, cloudiness and destruction of walls are the finding when the sinus is involved.
- *Eggshell of bone*—expansion and thinning of cortical plate occurs leaving thin 'eggshell of bone'. Perforation of bone is a late feature.
- *CT features*—computed tomography can explain exact nature and dimension of the lesion.
- *Radiological diagnosis*—honeycomb appearance, soap bubble appearance seen in posterior mandible with bony expansion.
- *Laboratory diagnosis*—in biopsy various types of ameloblastoma can be seen. It can be follicular (small discrete islands of tumor cells with peripheral layer of cuboidal and columnar ameloblast-like cells), plexiform type (ameloblasts-like tumor cells arranged in irregular masses or more frequently, as network of interconnecting strands of cells, each of which is bounded by layer of columnar cell and between these layers found stellate reticulum-like cells), Acanthomatous type (cells occupying the position of stellate reticulum undergo squamous metaplasia, with keratin formation in the central portion of tumor islands), granular (stellate reticulum cells takes coarse granular eosinophilic appearance), basal type (show baseloid pattern), unicystic (same like dentigerous cyst with ameloblastic changes).

Diagnosis

- *Clinical diagnosis*—swelling in posterior mandible with expansion and eggshell cracking will give clue to diagnosis.

Types of Ameloblastoma

Peripheral Ameloblastoma

It is also called as 'extraosseous ameloblastoma'. It is a tumor which occurs in the soft tissue outside and overlying the alveolar bone. It originates from either the surface epithelium or the remnants of dental lamina.

Clinical Features

- **Age and sex distribution**—it is common in males and more commonly found in middle age group.
- **Site**—it is seen on the mandibular gingiva in the molar-premolar region. Some cases also seen in buccal mucosa and maxillary tuberosity area.
- **Symptoms**—the patient complains of an intraoral mass which is growing in size and interfering with speech, mastication and esthetics.
- **Sign**—mass is mildly tender on palpation when ulcerated and bleeding may occur on slight touch.
- **Appearance**—The lesion appears nodular swelling on gingiva varying in size from 3 mm to 2 cm in diameter.

Radiological Features

- **Bone erosion**—sometimes underlying bone may exhibit evidence of pressure resorption in the form of saucer-shaped depression beneath the tumor.

Diagnosis

- **Clinical diagnosis**—not so specific.
- **Radiological diagnosis**—saucer shaped bone depression beneath the tumor mass may suspect the lesion.
- **Laboratory diagnosis**—biopsy shows same pattern as seen in intraosseous ameloblastoma.

Management

- **Surgery**—simple surgical excision of the lesion is treatment of choice. Prognosis of this tumor is very good.

Pituitary Ameloblastoma

It is also called as 'craniopharyngioma' or 'Rathke's pouch tumor'.

- **Origin**—it occurs in anterior lobe, which is of ectodermal origin. The lobe is derived from Rathke's pouch, an outgrowth of oral ectoderm. The pouch gives rise to craniopharyngeal duct, which in due course degenerates itself leaving residues of squamous epithelial cells. From these squamous epithelial cells, ameloblastoma-like tumor develops.
- **Age**—it is most common in childhood and adolescents before the age of 25 years.

- **Location**—it may be located within the sella tursica or commonly in suprasellar space. It grows as a pseudo-encapsulated mass causing pressure effect and often destroys the pituitary gland.
- **Symptoms**—they are related to destruction of pituitary gland (diabetes insipidus) or to compression of nearby cranial nerve.

Adamantinoma of Long Bones

- **Origin**—it is derived from epithelial rests misplaced during the course of development. Trauma causing implantation of epithelium with subsequent tumor formation.
- **Site**—it occurs in tibia, ulna, femur and occasionally in others bone.
- **Clinical course**—some lesions behave aggressively and sometimes are metastasizing.

Differential Diagnosis (Radiological)

Small and unilocular ameloblastoma

- **Residual cyst**—there is history of extraction of the teeth.
- **Lateral periodontal cyst**—found in incisor, canine and premolar area in maxilla and ameloblastoma occur in mandibular molar area.
- **Giant cell granuloma**—anterior to the molar and ameloblastoma occur in molar area.
- **Traumatic bone cyst**—occurs in mid-twenties and ameloblastoma common in 3rd and 4th decade.
- **Primordial cyst**—same as traumatic bone cyst.

Multilocular ameloblastoma

- **Odontogenic myxoma**—history of missing tooth and has a presence of septa that divide the image into much finer course than those in ameloblastoma.

Management

- **Curettage**—it is least efficient as it makes recurrence inevitable. Curettage may leave bone that is invaded by tumor cells.
- **Intraoral block excision**—excision of a block of bone may be recommended if the ameloblastoma is small. The segment in which the tumor is contained is removed with a safe margin of normal bone.
- **Extraoral En-block resection**—if the lesion is large and horizontal ramus is involved then this is carried out.
- **Peripheral osteotomy**—it is a procedure which allows complete excision of the tumor but at the same time a part of bone is retained which preserves the continuity of the jaw. The procedure is based on the observation that cortical inferior border of the horizontal body, the posterior border of the ascending ramus and condyle are generally not involved by tumor process. These areas are resistant and strong because of the dense cortical bone. Bone regeneration will proceed from such areas with considerable restoration of the jaw architecture.

Squamous Odontogenic Tumor

It is well differentiated odontogenic tumor composed of islands or sheets of squamous epithelium that lack recognizable features of enamel organ differentiation. It arises from the cell rests of Malassez, although hamartomatous epithelial transformation is also found.

Clinical Features

- *Age and sex*—there is a wide age distribution in the range of 11 to 67 years with mean age 40 years and females are more commonly affected than the males.
- *Site*—it occurs with equal frequency in maxilla and mandible. In maxilla, it occurs in incisor-cuspid area and in the mandible; it has got a predilection for the bicuspid-molar area.
- *Onset*—it is slowly growing tumor.
- *Symptoms*—it is usually asymptomatic but there may be mobility of the involved teeth, pain, tenderness to percussion and occasionally abnormal sensation.

Radiological Features

- *Site*—it occurs usually in association with the cervical portion of the tooth.
- *Radiodensity*—it is well circumscribed radiolucent area.
- *Shape*—it presents as a semicircular or roughly triangular area (**Fig. 14-24**).
- *Margin*—border may or may not be sclerotic.

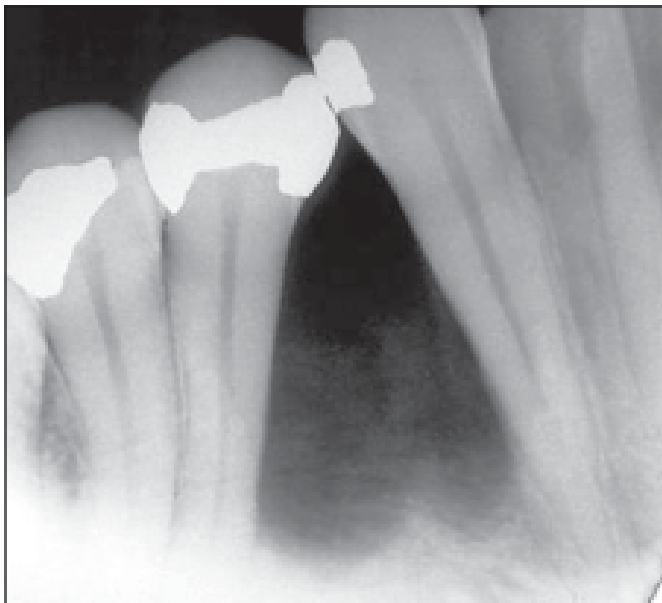


Fig. 14-24: Semicircular shape radiolucency seen squamous odontogenic tumor.

Diagnosis

- *Clinical diagnosis*—not so specific.

- *Radiological diagnosis*—semicircular radiolucency present between the roots.
- *Laboratory diagnosis*—biopsy shows islands of squamous epithelium without peripheral palisaded or polarized columnar cells.

Differential Diagnosis

It is a rare lesion so, it should not be high on the differential diagnostic list when considering radiolucent jaw lesions. If at exploration one discovers a solid fleshy lesion which is associated with a vital tooth root on cervical portion then squamous odontogenic tumor is the most likely diagnosis.

Management

- *Surgery*—conservative enucleation and curettage is usually curative with a low recurrence rate.

Odontogenic Myxoma

It is also called as 'odontogenic fibromyxoma', 'myxofibroma'. It is a rare non-invasive neoplasm that arises from the dental papilla, follicular mesenchyme and periodontal ligament.

Origin

- *Overgrowth of dental papilla*—the myxomatous tissue arises as a direct outgrowth of dental papilla of tooth or as an indirect effect of odontogenic epithelium on mesenchymal tissue.
- *Changes in fibrous tissue*—as a direct myxomatous change in fibrous tissue.

Clinical Features

- *Age and sex*—it is slightly more common in females with an age range of 10 to 30 years.
- *Incidence*—it accounts for 3 to 6% of odontogenic tumors.
- *Site*—mandible is more commonly affected than maxilla by a ratio of 3:1. Premolar-molar area in mandible and zygoma in maxilla. Rarely tumor may appear in the condylar region.
- *Symptoms*—they are associated with congenitally missing teeth. The growth rate is slow and pain is variable. There is a hard swelling which may be sometime large enough to produce facial asymmetry.
- *Signs*—sometimes it perforates the cortical plate producing a bosselated surface (several small nodules on the surface).
- *Appearance*—it may appear as fusiform swelling that may be hard and or may be covered by a layer of bone of only eggshell.

- **Teeth**—it may cause expansion and become huge if unattended. Teeth will be displaced and loosened but root resorption is rare.
- **Maxillary sinus**—it can invade maxillary sinus and cause exophthalmos.

Radiographic Features

- **Internal structure**—it may be either unilocular or multilocular. It may be mixed radiopaque-radiolucent lesion. Compartments tend to be angular. They may be separated by straight septa that form square, rectangular or triangular spaces. The central portion is transversed by fine gracile trabeculation.
- **Margins**—it is usually well defined but sometimes it may be poorly defined. It may be scalloped between the roots of adjacent teeth.
- **Tennis racket or honeycomb appearance**—locules are small and uniform with typical honeycomb appearance or strings of tennis racket (**Fig. 14-25**). Trabecular are arranged in right angle to each other.
- **Soap bubble appearance**—exceptionally, fine septa crosses the radiolucent area producing a soap bubble appearance.
- **Roots**—roots rarely show resorption.
- **Margin**—some of the myxomas resemble a dental cyst and instead of the smooth borders seen with the cyst, there are crenated margins.

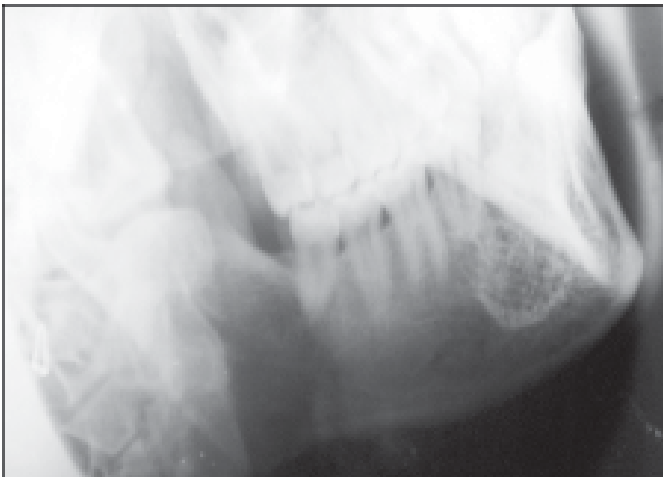


Fig. 14-25: Tennis racket pattern seen in odontogenic myxoma.

Diagnosis

- **Clinical diagnosis**—missing teeth with hard swelling may suspect this disease.
- **Radiological diagnosis**—tennis racket pattern seen in odontogenic myxoma.
- **Laboratory diagnosis**—biopsy shows loosely arranged, spindle shaped and stellate cells, many of which have long fibrillar processes that tend to be intermeshed.

Differential Diagnosis

- **Central giant cell granuloma**—anterior location in mandible.
- **Ameloblastoma**—usually occurs in older patients.
- **Cherubism**—younger age group and bilateral involvement.
- **Giant cell lesions of hyperthyroidism**—history of kidney disease and abnormal serum chemistry.
- **Metastatic carcinoma**—older age group and presence of primary tumor.
- **Aneurysmal bone cyst**—it is tender and painful.
- **Central hemangioma**—it is not associated with missing tooth. Pumping tooth syndrome is seen in hemangioma with aspiration being useful.

Management

- **Surgical excision**—tumors may be difficult to enucleate due to their loose consistency, therefore surgical excision is indicated.
- **Resection**—resection with generous amount of surrounding bone.

Peripheral Odontogenic Fibroma

It is also called as 'peripheral ossifying fibroma', 'peripheral ameloblastic fibrodentinitoma', 'calcifying fibrous epulis' 'peripheral fibroma with calcification'. It consists of fibrous tissue containing nests of odontogenic epithelium and calcified material that resembles cementum.

It involves periodontal ligament superficially and contains odontogenic epithelial nests and deposits of cementum, bone and dystrophic calcifications scattered throughout the background of fibrous tissue.

Etiology

- **Irritation**—it is associated with irritation (overextended margin of faulty restoration and deposits of calculus).

Clinical Features

- **Sex and age distribution**—age 5 to 25 with peak at 13 years. Females are affected more than males.
- **Common sites**—it occurs more commonly in the mandible. It occurs exclusively on free margin of the gingiva and involves interdental papillae.
- **Symptoms**—it is usually asymptomatic. It is slow growing, often present for a number of years. Some lesions grow large enough to cause facial asymmetry.
- **Signs**—they are solid and firmly attached to the gingival mass, sometimes arise between teeth and sometimes displaces the teeth (**Fig. 14-26**).

- **Appearance**—it is well demarcated mass of tissue on the gingiva with a sessile or pedunculated base. Early lesions are soft, quite vascular and red and bleed readily. More mature lesions are firm, fibrous and pale pink.



Fig. 14-26: Peripheral odontogenic fibroma presenting as mass attached to gingiva (Courtesy Dr Abhishek Soni).

Radiographic Features

- **Bone erosion**—on rare occasion, there may be superficial erosion of bone.
- **Calcification**—there may be presence of some radiopaque foci within the tumor mass which occur due to calcifications within the tumor.

Diagnosis

- **Clinical diagnosis**—sessile mass attached to gingiva may suspect peripheral odontogenic fibroma.
- **Radiological diagnosis**—not specific.
- **Laboratory diagnosis**—biopsy shows cellular fibrous connective tissue parenchyma with non-neoplastic islands, strands and cords of columnar or cuboidal cells.

Differential Diagnosis

- **Chondrosarcoma and osteogenic sarcoma**—less frequent gingival lesion, severe bony changes, asymptomatic with widening of PDL.
- **Inflammatory hyperplasia**—no separation of teeth occurs.

Management

- **Surgical excision**—simple surgical excision and submitted for microscopic examination for confirmation of diagnosis.

Central Odontogenic Fibroma

It is a lesion around the crown of an unerupted tooth resembling a small dentigerous cyst. But some say that

it is a hyperplastic dental follicle and not an odontogenic tumor. It may occur centrally or in the periphery.

Clinical Features

- **Age and sex distribution**—it occurs more frequently in older individual with mean age of 40 years. There is marked female predilection.
- **Site**—it is more common in maxilla and in anterior region.
- **Symptoms**—it is generally asymptomatic except for the swelling of the jaws.
- **Signs**—it may cause localized bony expansion and loosening of teeth.

Radiological Features

- **Radiodensity**—it produces an expansile radiolucency similar to that of ameloblastoma.
- **Site**—it is often associated with apices of erupted teeth.
- **Margin**—margins are well defined and sclerotic.
- **Internal structure**—it can be unilocular but larger lesions tend to be multilocular.
- **Effect on surrounding structures**—larger lesions may cause root divergence and resorption.

Diagnosis

- **Clinical diagnosis**—not so specific.
- **Radiological diagnosis**—multilocular radiolucency in anterior maxilla may suspect central odontogenic fibroma (**Fig. 14-27**).
- **Laboratory diagnosis**—biopsy shows tumor mass made up of mature collagen fibers interspersed usually by many plumps of fibroblasts that are very uniform in their placement and tend to be equidistant from each other.

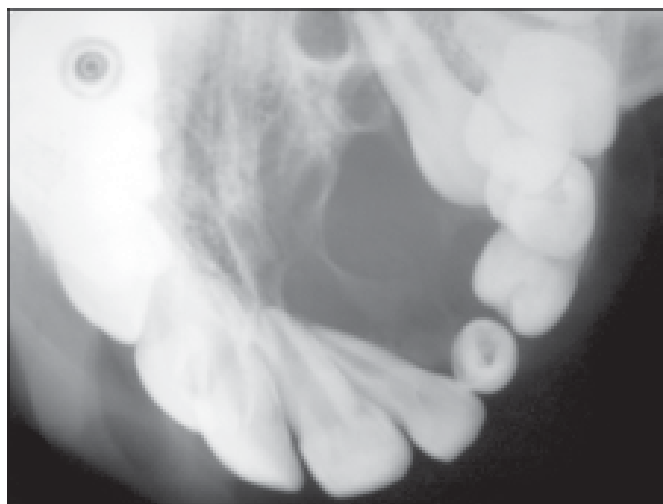


Fig. 14-27: Multilocular appearance of central odontogenic fibroma present in anterior maxilla.

Management

- *Curettage and enucleation*—central odontogenic fibroma is treated with enucleation and curettage. It does not recur.

Granular Cell Odontogenic Tumor

It is also called as '*granular cell odontogenic fibroma*'. It is more cellular. It consists of large eosinophilic granular cells with cords and islands of odontogenic epithelium.

Clinical Features

- *Age and sex distribution*—it occurs more frequently in older person (more than 40 years of age). There is slightly female predilection.
- *Site*—it is common in premolar and molar area of mandible.
- *Symptoms and sign*—lesion is painless with expansion of the jaw is noted.

Radiological Features

- *Appearance*—it appears as well demarcated radiolucency. It is unilocular or multilocular.
- *Calcification*—small degree of calcification is noted in the lesion.

Diagnosis

- *Clinical diagnosis*—not specific.
- *Radiological diagnosis*—calcification in unilocular radiolucency may suspect the lesion.
- *Laboratory diagnosis*—biopsy shows large eosinophilic granular cells. Narrow cords or small islands of odontogenic epithelium are scattered in granular cells area. Small cementum-like calcification is also seen.

Management

- *Curettage*—simple curettage of the lesion should be carried out. No recurrence is reported.

Periapical Cemental Dysplasia

It is also called as '*fibrocementoma*', '*sclerosing cementum*', '*periapical osteofibrosis*', '*periapical fibrosarcoma*'. It is a reactive fibro-osseous lesion derived from the odontogenic cells in the periodontal ligament. It is located at the apex of the teeth. It is usually discovered as an incidental finding during routine radiographic surveys.

Etiology

- *Local factors*—it occurs as a result of trauma, chronic irritation.

- *Hormone*—there is possibility that female sex hormones may be an etiologic factor since the disease is commonly found in women.
- *Others*—nutritional deficiency, metabolic disturbances, past history of syphilis, endocrinal imbalance and anomalous development.

Pathogenesis

Bone at the apex of the affected tooth may be replaced by fibrous tissue which may be small in amount or occupy an area of a square inch or more →the fibrous tissue may remain unchanged for months to years but in most instances, there is a tendency for a change to occur →the affected area may become partially reduced in size by peripheral ossification →bone or cementum forms either as a single mass which enlarges by aggregation at its margin or as several masses →this mass either enlarges individually or they fuse →the fibrous tissue may be replaced entirely by bone of greater density than that of normal bone →once cementum is formed it remains unchanged without any reduction.

Clinical Features

- *Age and sex distribution*—it occurs during the middle age with a mean age of 39 years. Male to female ratio is 1:9 and is three times more common in blacks than in whites.
- *Site*—mandibular anterior region is commonly affected. Single area of one jaw may be involved or the greater part or both the jaws is affected.
- *Symptoms*—involved teeth are vital with no history of pain or sensitivity. Occasional lesions localize near the mental foramen and impinge on the mental nerve and produce pain, paresthesia or even anesthesia.
- *Signs*—hypercementosis is usually associated with it. It rarely enlarges.

Radiographic Features

- *Site*—it usually lies at the apex of the tooth. In rare cases, epicenter is high and over the apical thirds of the root. In most cases, lesion is multiple and bilateral.
- *Margins*—margins are well defined. A radiolucent border of varying width, surrounded by band of sclerotic bone is seen. Sclerotic bone represents immediate reaction of surrounding bone.
- *Stage I—Radiolucent (fibrous)*
 - *Radiodensity*—since there is loss of bony substance and replacement by connective tissue, the lesions appear radiolucent.
 - *Shape*—there is formation of a circumscribed area of periapical fibrosis accompanied by localized bone destruction (Fig. 14-28).

- *Margins*—the margins of the radiolucent area vary in different lesions; some are well defined but not corticated and others are poorly defined or well defined at one portion of the lesion and ill defined elsewhere. A rim of slightly denser bone surrounds the greater part or all the periphery of some lesion.
- *Lamina dura*—lamina dura around the tooth is lost.



Fig. 14-28: Early stage of PCD showing radiolucency at the periapical area of mandibular incisor.

- *Stage II—Mixed stage*
 - *Margins*—margins of the opacity are usually sharp but sometimes it can be ill defined and irregular.
 - *Internal structure*—minute radiopacities are seen within the radiolucent periapical area due to either bone or cementum formation (**Fig. 14-29**). It is called as *cementoblastic stage*. Small radiopacities may coalesce.
 - *Size*—opacity may vary from a millimeter or a centimeter or more.
 - *Shape*—it has round, oval or irregular shape.
- *Stage III—Radiopaque*
 - *Radiodensity*—there is complete opacification (**Fig. 14-30**).
 - *Appearance*—it appears as a well defined radiopacity usually bordered by a radiolucent capsule separating it from the adjacent bone.
 - *Margins*—margins may vary from well defined to poorly define.
 - *Lamina dura*—lamina dura of adjacent teeth may be discontinuous in the area of lesion.
 - *Hypercementosis*—hypercementosis is produced in some cases which can be seen at the apex.
 - *Effect on surrounding structures*—root resorption is rare. The lesion can elevate the floor of maxillary antrum.



Fig. 14-29: Mixed stage of PCD showing radiopacity in the radiolucency.

- Some of these large dense areas reveal small dark spot which resemble dense bone with persistent small marrow spaces.



Fig. 14-30: Mature stage of PCD showing complete radiopaque lesion.

Diagnosis

- *Clinical diagnosis*—not so specific.
- *Radiological diagnosis*—radiolucency at the apex of vital teeth with no loss of lamina dura in early stage. In the mature stage radiopaque lesion is seen.

- *Laboratory diagnosis*—biopsy shows periapical bone is replaced by mass of fibrous tissue. Small, round to ovoid calcifications are deposited within the fibrous tissue.

Differential Diagnosis

The osteolytic stage or early stage

- *Pulpo-periapical lesions*—are associated with pulpal disease and the involved tooth is sensitive to percussion. Any radiolucent area at the apex of the tooth, particularly lower incisor, which does not show caries or restoration, would be suspected periapical cemental dysplasia.
- *A traumatic bone cyst*—when projected over a tooth with a vital pulp can be confused with periapical cemental dysplasia; but it is usually much larger and is characteristically found in a younger age group.
- *The cementifying and ossifying fibroma in early stages*—it also occurs at the apex of vital teeth in early stages but it affects a younger age group and they have a potential to be a very large lesions. They do not have a predilection for the lower incisor area but occurs most often in the premolar region.
- *Cementoblastoma*—it can be confused in early stages but it is a rare lesion that occurs almost exclusively at the apex of mandibular molars. It extends higher on the root. It is connected to the root surface in mandibular premolar and molar area.

Intermediate stage/mixed stage

- *Malignant osteoblastic carcinoma*—it is rapidly growing with borders which are irregular and ill defined.
- *The intermediate stage of odontoma*—it shows orderly relationship of radiopaque enamel, dentin and pulp space in compound odontoma; while in cases of complex odontoma, more radiopaque component of enamel will give a clue towards the true identity of the lesion. In addition, odontoma is located above the crown of an unerupted tooth, sometimes between teeth, but rarely in the periapical region.
- *Calcifying crown*—it presents in the 1st and 2nd decade and is easily identified by their anticipated location in the jaw and the presence of a similar picture in the contralateral jaw.
- *Fibrous dysplasia*—it occurs in a younger age group (10-20 years) than periapical cemental dysplasia and is common in the maxilla. Margins are poorly defined in fibrous dysplasia and well defined in periapical cemental dysplasia.
- *Periapical rarefying osteitis*—is seen with non-vital teeth. Absence of pain, tenderness on palpation, inflammation and no regional lymphadenopathy favors the diagnosis of periapical cemental dysplasia.
- *Cementifying and ossifying fibroma*—periapical cemental dysplasia is a common lesion as compared to

cementifying and ossifying fibroma. Periapical cemental dysplasia has a predilection for lower incisors while cementifying and ossifying fibroma is common in the premolar- molar region. Periapical cemental dysplasia occurs in a younger age group as compared to cementifying and ossifying fibroma. Periapical cemental dysplasia seldom attains a diameter above 1 cm while cementifying and ossifying fibroma attains a diameter of 2-4 centimeters.

Mature stage/radiopaque

- *Hypercementosis*—it involves anterior teeth and is attached to a part of the root and is separated from periapical bone by the radiolucent periodontal ligament space which surrounds the entire root.
- *Condensing osteitis*—it occurs at the apices of non-vital teeth whereas periapical cemental dysplasia does not. In addition, condensing osteitis does not have a radiolucent rim whereas it is present in periapical cemental dysplasia.
- *Periapical idiopathic osteosclerosis*—it may be difficult to differentiate because both occur in the periapex of healthy teeth with vital pulp. Periapical cemental dysplasia is smoothly contoured and almost always round or ovoid whereas periapical idiopathic osteosclerosis is usually quite irregular in shape and also there is absence of the radiolucent rim.
- *Paget's disease and osteoblastic metastatic carcinoma*—can be ruled out if there is absence of systemic symptoms.

Management

- *Continuous observation*—it requires continuous observation and subsequent verification through periodic radiographic examination.
- *Surgical enucleation*—surgical enucleation is indicated for larger lesions which have caused expansion of the cortical plates or when the clinician is unsure of the working diagnosis.

Benign Cementoblastoma

The benign cementoblastoma is also called as *true cementoma*. It is rare odontogenic tumor representing less than 1% of all odontogenic tumors.

Norberg initially described it in 1930 and defined as a true neoplasm of cementum or cementum-like tissue that formed on a tooth root by cementoblasts.

The WHO defines the benign cementoblastoma as 'a neoplasm' characterized by the formation of sheds of cementum-like tissue which may contain a very large number of reversal line and may be unmineralized at the periphery of the mass or in the more active growth areas.

The accepted theory of its origin is that it is a mesenchymal odontogenic tumor. The cementoblastoma's

precise derivation is connective tissue of the periodontal ligament.

Clinical Features

- *Age and sex distribution*—the benign cementoblastoma occurs most frequently under the age of 25 years and there appears to be a slight predilection for males.
- *Site*—cementoblastoma can occur in both maxilla and the mandible. The mandible, however, is involved three times more frequently than maxilla. The mandibular first permanent molar is the most frequently affected tooth.
- *Symptoms*—pain is frequently present and it is the most common symptom.
- *Sign*—the associated tooth is vital unless coincidentally involved. The lesion is slow growing and produces cortical expansion. Extraoral (**Fig. 14-31**) and intraoral swelling can also be present (**Fig. 14-32**).



Fig. 14-31: Swelling of the jaw seen extraorally due to cementoblastoma



Fig. 14-32: Cementoblastoma intraoral view showing intraoral swelling.

Radiographic Features

- *Margins*—well defined radiopacities usually attached to the roots of premolars and molars surrounded at the border by a radiolucent halo are seen.
- *Appearance*—the radiological appearance of the cementoblastoma is highly characteristic, seen as circular radiopaque mass attached to the root of the one or more teeth. A narrow radiolucent zone surrounds the lesion and delineates from adjacent bone (**Fig. 14-33**).
- *Root outline*—the outline of the affected root is generally obliterated because of resorption of the root and fusion of the mass to the tooth.
- *Internal structure*—they are mixed radiolucent-radiopaque lesion that may be amorphous or may have *wheel-like spoke pattern*. Density of cementum obscures the outline of the enveloped root.
- *Effect on surrounding structures*—the outline of the tooth is obliterated because of resorption of the root and fusion of the mass to the tooth.
- *Expansion*—occlusal radiograph will demonstrate its expansive nature.

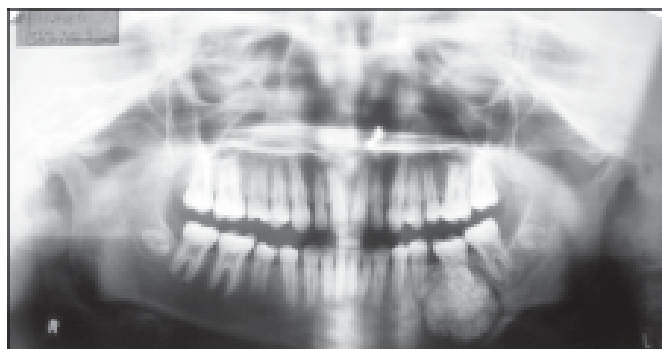


Fig. 14-33: Benign cementoblastoma showing radiopacity surrounded by radiolucent capsule.

Diagnosis

- *Clinical diagnosis*—not so specific
- *Radiological diagnosis*—radiopaque lesion surrounded by radiolucent capsule attached to root surface is typical of cementoblastoma.
- *Laboratory diagnosis*—biopsy shows cementum-like tissue, deposited in globular pattern resembling giant cementicles.

Differential Diagnosis

- *Periapical cemental dysplasia*—there is no expansion of the jaws and females are more commonly affected.
- *Chronic focal sclerosing osteomyelitis*—no radiolucent halo is present.
- *Periapical osteosclerosis*—radiolucent halo present in case of cementoblastoma and it is absent in periapical osteosclerosis.

- *Hypercementosis*—cementoblastoma initially associated with larger root than hypercementosis and is surrounded by radiolucent halo.

Management

- *Excision*—the recommended treatment of cementoblastoma usually consists of the surgical extraction of the tooth together with the attached calcified mass. Surgical excision of the mass with root amputation and endodontic treatment of the involved tooth may be considered. The prognosis is excellent and the tumor does not recur after total removal. Recurrence is not seen.
- *Endodontic treatment*—tumor can be amputated from the tooth and tooth is then endodontically treated.

Ameloblastic Fibro-odontoma

The name is applied to an odontogenic lesion that exhibits features of ameloblastic fibroma as well as dentin and enamel. It is mixed epithelial-mesenchymal proliferation and both mature and immature areas.

Clinical Features

- *Age and sex*—it is usually seen in a young age group with mean age of occurrence of 12 years. Males are more commonly affected than females.
- *Site*—maxilla and mandible are equally affected. In mandible, it occurs in the molar area and in the maxilla it usually involves the maxillary sinus.
- *Symptoms*—the most common presenting complaint is swelling and failure of tooth eruption. The maxillary tumor if large interferes with nasal respiration, eating and speech.
- *Signs*—ameloblastic fibro-odontoma consists of elements of ameloblastic odontoma and is more aggressive than the common odontoma.

Radiological Features

- *Radiodensity*—a well-circumscribed lesion presenting as an expansile radiolucency generally containing either a solitary radiopaque mass or multiple small opacities representing the odontoma portion of the lesion (**Fig. 14-34**).
- *Size*—some of the lesions are relatively small, not over 1 to 2 cm in diameter; while others may be exceedingly large involving a considerable portion of the body of mandible and extending in the ramus.

Diagnosis

- *Clinical diagnosis*—not so specific.
- *Radiological diagnosis*—radiopaque calcified mass in the radiolucent defect.

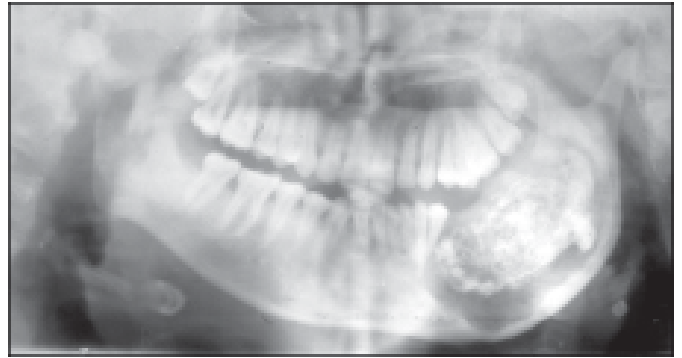


Fig. 14-34: Radiolucent defect showing radiopaque structure representing ameloblastic fibro-odontoma (Courtesy Dr Parate).

- *Laboratory diagnosis*—biopsy shows epithelial component with dysplastic dentin. There are cords, fingers, strands and rosettes of primitive odontogenic columnar or cuboidal epithelial cells often resembling dental lamina. The mesenchymal component is an embryonic fibrous connective tissue with delicate fibrils interspersed by large primitive fibroblast all resembling dental papilla.

Management

- *Curettage*—it is treated by curettage since it does not appear to locally invade the bone.

Ameloblastic Odontoma

It is also called as 'odontoameloblastoma'. It is characterized by the simultaneous occurrence of an ameloblastoma and a composite odontome. It is extremely rare odontogenic tumor.

Clinical Features

- *Age*—it occurs in children early in the second decade of life.
- *Site*—greater incidence in mandible as compared to maxilla.
- *Symptoms*—it produces considerable facial deformity or asymmetry if left untreated. Mild pain may be present as well as delayed eruption of teeth also occurs.
- *Sign*—it causes bony expansion and destruction of the cortex and displacement of teeth.

Radiological Features

- *Radiodensity*—radiographic density of ameloblastic odontoma may be radiopaque and similar to the complex odontoma or may be mixed.
- *Internal structure*—there is presence of numerous small radiopaque masses (**Fig. 14-35**) which may or may not bear resemblance to the formed albeit miniature teeth. In

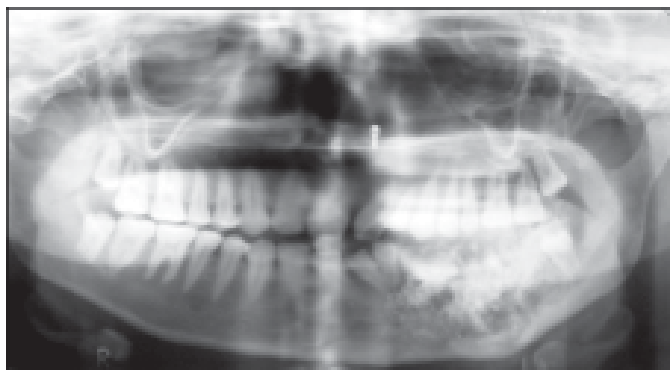


Fig. 14-35: Large masses of calcified structure seen in ameloblastic odontoma (Courtesy Dr Parate).

some cases, there is only a single irregular radiopaque mass of calcified tissue present.

- **Margins**—are well defined, uniformly smooth and with even border.
- **Cortical plate**—central destruction of bone with expansion of the cortical plates is prominent.
- **Effect on surrounding structures**—it can cause bone expansion, destruction and tooth displacement may occur.

Diagnosis

- **Clinical diagnosis**—not so specific.
- **Radiological diagnosis**—radiopaque calcified mass with cortical destruction.
- **Laboratory diagnosis**—biopsy shows undifferentiated epithelial cells with ameloblasts, enamel and enamel matrix, dentin, osteodentin, dentinoid and osteoid material. There is presence of sheets of typical ameloblastoma of one or the other of the recognized types usually basal cells, follicular or plexiform cells.

Management

- **Same as that of ameloblastoma**—tumor appears to have the same recurrence potential as an ameloblastoma and therefore should be treated similarly.

Malignant Tumor

Ameloblastic Fibrosarcoma

It is the malignant counterpart of the ameloblastic fibroma in which mesenchymal elements has become malignant. It is also called as 'ameloblastic sarcoma'.

Clinical Features

- **Age and sex**—it is very rare and most frequently occurs in young adults with an average age of occurrence being

30 years. It is more commonly seen in males as compared to female.

- **Site**—it occurs more frequently in mandible as compared to maxilla.
- **Symptoms**—it is uniformly painful, generally grows readily and causes destruction of bone with loosening of teeth.
- **Sign**—there may be ulceration and bleeding of the overlying mucosa. Swelling is usually soft in consistency.

Radiological Features

- **Radiodensity**—there is severe bone destruction (**Fig. 14-36**).
- **Margins**—it has irregular and poorly defined margins.
- **Effect on cortical bone**—there may be gross expansion and thinning of cortical bone.
- **Maxillary lesion**—in maxillary lesions, the involvement of the antrum may occur.



Fig. 14-36: Ameloblastic fibrosarcoma showing severe bone destruction.

Diagnosis

- **Clinical diagnosis**—not so specific.
- **Radiological diagnosis**—irregular bone destruction with ill defined margin can give clue to the diagnosis.
- **Laboratory diagnosis**—mesenchymal tissue exhibits a remarkable increase in cellularity; the malignant fibroblast being bizarre and pleomorphic with hyperchromatic nuclei and numerous atypical mitotic figures.

Management

- **Radical resection**—hemimandibulectomy or hemimaxillectomy can be done in patient of ameloblastic fibrosarcoma. Recurrence is common and prognosis is poor.

Ameloblastic Odontosarcoma

It is similar to ameloblastic fibrosarcoma but limited amounts of dysplastic dentin and enamel are formed.

Malignant Ameloblastoma and Ameloblastic Carcinoma

It is a rare type of tumor and diagnosis depends upon presence of metastases, which in some cases is seen in lymph nodes and lungs.

Terminology

- *Malignant ameloblastoma*—are those ameloblastomas that metastasize but in which the metastatic lesion do not show any histological difference from the primary tumor.
- *Ameloblastic carcinoma*—shows obvious histological malignant transformation but the metastatic lesions do not bear resemblance to the primary odontogenic tumor.

Clinical Features

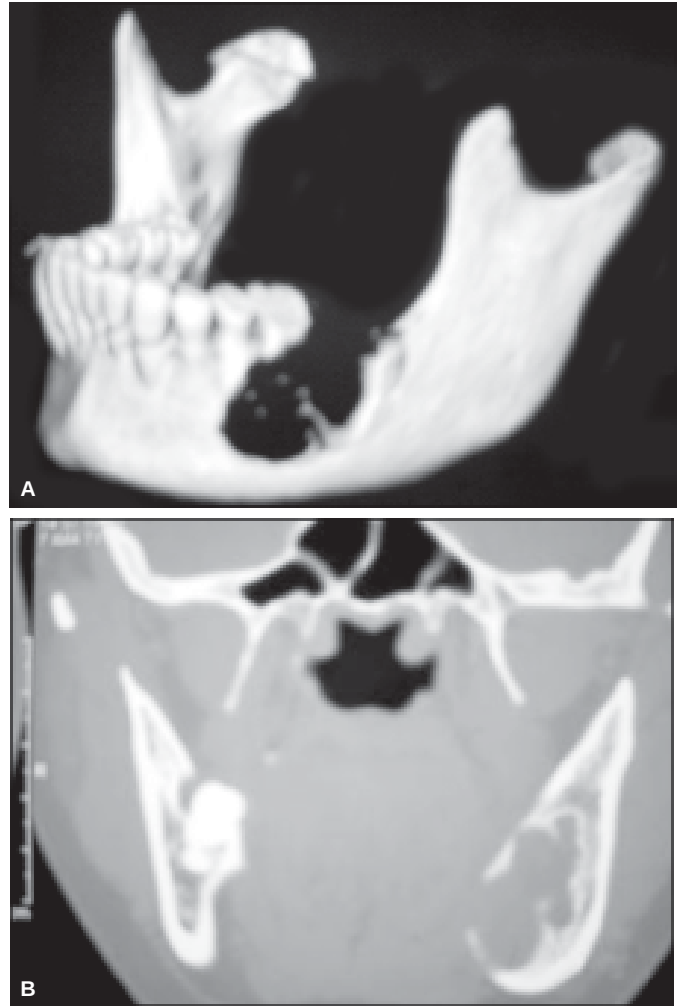
- *Age and sex*—males are affected more commonly than females. It occurs in 1st to 6th decade with mean age of diagnosis 28-32 years.
- *Site*—it is almost exclusively seen in the mandible.
- *Symptoms*—swelling followed by pain and/or rapid growth.
- *Sign*—teeth may be displaced and loosened. Tenderness of overlying soft tissue is present.
- *Sites for metastasis*—lungs, spleen, kidney, lymph nodes and ileum.
- *Local extension*—it may occur in adjacent bone, connective tissue or salivary gland.

Radiological Features

- *Site*—it is more common in premolar-molar area of mandible.
- *Margin*—it has well defined border with cortication, presence of crenations or scalloping in the perimeter. There may be loss of cortical boundary and breaching of the cortical boundary with soft tissue spread (**Figs 14.37A and B**).
- *Appearance*—it is either unilocular or multilocular giving the appearance of honeycomb or soap bubble pattern. Most of the septa are robust and thick.
- *Effect on surrounding structure*—teeth may be displaced and may exhibit root resorption. Lamina dura may be lost. Bony borders may be effaced or breached. The mandibular neurovascular canal may be displaced or eroded.

Differential Diagnosis

- *Benign ameloblastoma*—in it, there is no evidence of metastasis in benign ameloblastoma.
- *Central giant cell granuloma*—if the lesion is anterior to the premolar, central giant cell granuloma should be suspected.



Figs 14.37A and B: Malignant ameloblastoma showing loss of cortical boundary (Courtesy Dr Iswar).

- *Malignant changes in odontogenic cyst*—if the lesion is locally invasive and it is apparent radiologically then malignant changes in odontogenic cyst should be suspected.
- *Odontogenic myxoma*—there is missing tooth associated with it.

Diagnosis

- *Clinical diagnosis*—not so specific.
- *Radiological diagnosis*—loss of cortical boundary with features suggestive of ameloblastoma.
- *Laboratory diagnosis*—there may be cytological features of malignancy.

Management

- *En block resection*—it is treated with en bloc resection. Prognosis is poor in this case.
- *Pulmonary metastasis*—radiation therapy and chemotherapy for pulmonary metastasis.

Primary Intraosseous Carcinoma

It is also called as 'central mandibular carcinoma', 'primary intra-osseous carcinoma', 'primary epithelial tumor of the jaw', 'primary intra-alveolar epidermoid carcinoma' and 'central squamous cell carcinoma'. It develops within the depth of the jaw. It is rare and may remain silent until they have reached a fairly large size.

Origin

- *Malignant transformation*—primary intraosseous carcinoma may occur from malignant transformation of epithelial lining of odontogenic or non-odontogenic cyst, or ameloblastoma by metastases from different sites.
- *Primary tumor of maxillary sinus*—in case of maxilla, it may arise from primary tumor of the maxillary sinus.
- *Cell rests of odontogenic epithelium*—the tumor arises from the cell rests of the odontogenic epithelium or from the epithelial remnants at the site of fusion between two embryonic processes.
- *Cells of dental lamina*—in normal situation for some of the original cells of the dental lamina or enamel organ to remain in the jaw long after the function of these cells are completed. Malignant tumor may develop from these cells.

Types

- Those arising from odontogenic cysts.
- Those arising from ameloblastoma either well differentiated (malignant ameloblastoma) or poorly differentiated (ameloblastic carcinoma).
- Those arising de novo from odontogenic epithelium residues, either keratinizing or non-keratinizing.

Clinical Features

- *Age and sex distribution*—there is a wide range of age distribution with majority of cases occurring in 6th and 7th decade of life. It is more common in males than in females with a ratio of 2:1 and more common in mandible than in maxilla.
- *Symptoms*—the early symptom is swelling of the jaw with pain and mobility of the teeth before ulceration has occurred. Pathological fracture and lip paresthesia also occurs.
- *Signs*—there is rapid expansion and destruction of jaw bones. Tumor invades the periodontal ligament and the alveolar bone, destroying it. There may be lymphadenopathy. Surface epithelium is normal. Occasionally, the pulp of the teeth may be invaded by neoplasm. Perforation of cortical plate may occur.
- *Non-healing socket*—extraction of teeth result in non-healing socket and sometimes tumor may protrude from the non-healed socket.

Radiological Features

- *Radiodensity*—it presents as a diffuse radiolucency similar to other central malignant neoplasms of the jaws.
- *Site*—mandible is more commonly involved than the maxilla. Molar region is frequently involved than the anterior region.
- *Appearance*—central squamous cell carcinoma is very rare and appears as more or less rounded radiolucency completely surrounded by bone.
- *Margin*—its borders become ragged and there is no evidence of bone formation within the tumor.
- *Shape*—they are more often rounded or irregular in shape (Fig. 14-38). The purely lytic but irregular extension of the tumor may leave some areas of residual bone, either in the shape of islands or strands.

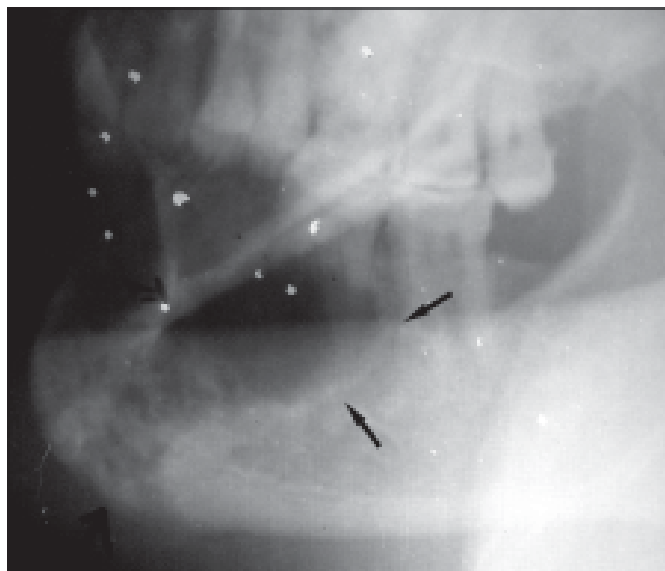


Fig. 14-38: Irregular radiolucency seen in mandible of intraosseous carcinoma.

- *Effect on surrounding structures*—there is expansion and distortion of the cortical plates of the jaw bone. They may cause destruction of the antral or nasal flows, loss of cortical outline of the mandibular neurovascular canal and effacement of the lamina dura.
- *Teeth*—teeth that lose both lamina dura and supporting bone appear to be floating in space.
- *Cortical plate and alveolar bone*—there may be perforation of the cortical plate and pathological fracture. There is severe destruction of alveolar bone with displacement of the teeth.

Diagnosis

- *Clinical diagnosis*—rapid expansion, lymphadenopathy, and pathological fracture may suspect this disease.
- *Radiological diagnosis*—it is purely lytic lesion with expansion and destruction of cortical plate.

- *Laboratory diagnosis*—biopsy shows plexiform pattern of peripheral cells of the tumor masses showing palisading arrangement, thereby resembling odontogenic epithelium. The tumor cells themselves generally exhibit nuclear pleomorphism and hyperchromatism, mitotic activity.

Differential Diagnosis

- *Periapical cyst or granulomas*—if the lesion is not aggressive, it may be mistaken for periapical cyst or granulomas. But in periapical cyst, tooth is non-vital and borders are well defined.
- *Odontogenic tumors*—margins are usually well defined as compared to carcinoma.
- *Metastatic carcinoma*—if the margins are obviously infiltrative with extensive bone destruction, it will be intraosseous carcinoma.
- *Peripheral squamous cell carcinoma*—surface epithelium is involved in peripheral squamous cell carcinoma.

Management

- *Surgical resection*—it is treatment of choice in primary intraosseous carcinoma.

Malignant Changes in Odontogenic Cyst

It is also called as '*carcinoma ex odontogenic cyst*'. It is uncommon lesion seen in oral cavity. There are carcinomatous changes found in odontogenic cyst. In some cases adjacent carcinoma may involve otherwise unrelated cyst. It occurs due to epithelial dysplasia occurring in the cyst lining.

Clinical Features

- *Age and sex distribution*—these are more commonly found in older patient with male predilection.
- *Site*—it can affect any cyst in the jaw. But keratocyst is more likely to undergo malignant changes than other cysts.
- *Symptoms*—the most common complaint is pain, which is dull and of several months duration. If upper jaw is involved, sinus pain and swelling may be present.
- *Sign*—pathological fracture, fistula formation and regional lymphadenopathy may occur.

Radiological Features

- *Site*—it is most commonly found in tooth bearing area of the jaw. Most commonly in the mandible.
- *Margin*—if it is small lesion in the cyst wall, the periphery may be well defined and corticated. As the malignant lesion progressively replaces cyst lining, the smooth border is lost or becomes ill defined. Advanced lesion

has an ill defined, infiltrative periphery that lacks any cortication.

- *Shape*—in initial lesion, the shape is ovoid or round. In advanced lesion, shape becomes less '*hydraulic*' looking and more diffuse.
- *Radiodensity*—it is more radiolucent than invasive surface carcinoma.
- *Effect on surrounding structure*—there is thinning and destruction of lamina dura of adjacent teeth. It can also destroy adjacent cortical boundaries such as inferior border of the jaw or floor of nose. It can produce complete destruction of the alveolar process.

Diagnosis

- *Clinical diagnosis*—not specific.
- *Radiological diagnosis*—smoothness of cyst is lost and margin becoming ill defined may give clue to the diagnosis.
- *Laboratory diagnosis*—they are same as that of squamous cell carcinoma.

Differential Diagnosis

- *Infected dental cyst*—if dental cyst is infected, it may lose its normal cortical boundary and appear to be ragged. But infected cyst usually shows a reactive peripheral sclerosis because of inflammatory products present in the cystic lumen.
- *Metastatic tumor*—it is multifocal and there is history of primary tumor.
- *Multiple myeloma*—multiple separate radiolucency can be seen.

Management

- *Radical resection*—treatment given is either local block excision or radical resection.

Odontogenic Fibrosarcoma

It is the malignant counterpart of odontogenic fibroma. It originates from same mesenchymal tissue, as same in central fibroma.

Clinical Features

It is a destructive lesion which produces a fleshy, bulky growth. It is asymptomatic but pain may be present in many cases.

Diagnosis

Biopsy shows cellular element that may or may not be prominent than the fibrillar component. The cells often exhibit considerable mitotic activity. They resemble imma-

ture fibroblasts and appear as elongated cells containing ovoid nuclei with varying degree of pleomorphism and are situated in a fibrous meshwork which may or may not exhibit foci of odontogenic epithelium.

Management

Radical surgical removal with resection of the jaw. Prognosis is poor.

Clear Cell Odontogenic Tumor or Carcinoma

It is rare odontogenic tumor with potential for lymphatic or pulmonary metastases. Its origin is unknown, but ultrastructural study shows clear cells, which have similarities to glycogen rich presecretory ameloblasts.

Clinical Features

- *Age and sex*—most cases reported in women over 60 years of age.
- *Site*—both maxilla and mandible have been involved.
- *Symptoms*—it may present as asymptomatic or painful bony swelling.

Radiological Features

- *Appearance*—unilocular or multilocular radiolucency with ill defined margins.

Diagnosis

- *Clinical diagnosis*—not specific.
- *Radiological diagnosis*—not specific.
- *Laboratory diagnosis*—biopsy shows nests of epithelial cells with clear or faintly eosinophilic cytoplasm which is separated by thin strands of hyalinized material.

Management

- *Extensive resection*—tumors demonstrate aggressive local behavior and potential lymphatic and pulmonary metastases and therefore should be treated with extensive resection.

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Introduction

It is a new growth representing the tissue of origin. The study of the tumors of the oral cavity and adjacent tissues constitute an important phase of dentistry because of the role of a dentist in the diagnosis of the lesion.

Characteristics of Benign Tumor

- *Insidious onset*—it has got insidious onset and the rate of growth is generally slow and is for a longer period of time.
- *Well defined mass*—it has got well defined mass of regular smooth outline and it possesses a fibrous capsule.
- *Symptoms*—benign tumors usually produce symptoms due to the swelling and pressure effect on the surrounding structures. It is usually painless and they never metastasize.
- *Small*—benign tumors are smaller, as compared to malignant tumors.
- *Displacement of teeth*—there is displacement of adjacent normal tissues.

Terminology

- *Hamartomas*—it is an abnormal proliferation of normal tissue at its usual location. For e.g. Hemangioma.
- *Neoplasm*—tumors that continue to grow indefinitely are called as neoplasm.
- *Hypertrophy*—enlargement caused by an increase in the size of cells.
- *Hypertrophy*—enlargement caused by an increase in the number of the cells.

Classification

Epithelial tissue

- Papilloma

- Keratoacanthoma
- Squamous acanthoma
- Nevus

Fibrous connective tissue

- Fibroma
- Fibrous hyperplasia
- Fibrous epulis
- Giant cell fibroma
- Fibrous histiocytomas
- Desmoplastic fibroma
- Myxoma
- Myxofibroma

Cartilage tissue

- Chondroma
- Chondroblastoma
- Chondromyxoid fibroma

Adipose tissue

- Lipoma
- Angiolipoma

Bone

- Osteoma
- Osteoid osteoma
- Osteoblastoma
- Exostosis-tori-torus palatinus-torus mandibularis
- Osteomatosis

Vascular tissue

- Hemangioma
- Hereditary hemorrhagic telangiectasia
- Lymphangioma
- Arteriovenous fistula
- Glomus tumor

Neural tissue

- Neurofibroma
- Neurolemmoma

- Schwannoma
- Ganglioneuroma
- Traumatic neuroma
- Melanotic neuroectodermal tumor of infancy

Muscles

- Leiomyoma
- Rhabdomyoma
- Granular cell myoblastoma

Giant cell tumor

- Central and peripheral giant cell tumor
- Giant cell granuloma
- Giant cell tumor of hyperthyroidism

Teratoma

Tumors of Epithelial Origin

Papilloma

It is relatively a common, benign neoplasm of unknown origin, which arises from the surface of stratified squamous epithelium. It may be caused by papilloma virus.

Types

- *Squamous cell papilloma*—it may arise in the tongue, cheek, lip and esophagus
 - *Congenital*—it is usually present since birth.
 - *Infective*—it arises from viral infection.
 - *Soft papilloma*—it is often seen in eyelids of elderly people.
 - *Keratin horns*—it is due to excess keratin formation and seen in older people.
- *Basal cell papilloma*—it is also called as 'seborrheic or senile wart'. It occurs on the trunk, face, arms and armpits.

Clinical Features

- *Age*—average age of occurrence is in the 3rd and 4th decade of life, only 20% of cases are found below 20 years of age.
- *Common sites*—it is most commonly seen on tongue, palate, buccal mucosa, gingiva, lip, mandibular ridge and floor of mouth.
- *Cauliflower appearance*—it is typically an exophytic lesion with a cauliflower-like surface or with finger-like projection (Fig. 15-1). Projections are pointed or blunt. This appearance is caused by presence of deep clefts that extend well into lesion from the surface.
- *Base*—it generally arises from a pedunculated base. Some time, base may be broad rather than pedunculated.
- *Shape*—small mass on mucosa with a papillomatous shape.

- *Size*—the size of tumor may vary from 2 millimeters but it is seldom larger than 2 centimeters.
- *Color*—tumor with much keratinization is white and lesion without much keratinization is grayish pink in color.
- *Consistency*—tumor is firm, when keratinized and soft when it is non-keratinized.
- *Focal dermal hypoplasia syndrome*—in it, there are multiple oral papillomas, dermal hypoplasia and syndactyly with fatty herniation, coloboma and strabismus.
- *Other syndromes associated*—other syndromes in which multiple papillomas may occur are Cowden's syndrome, Down's syndrome and nevus unius lateris syndrome.



Fig. 15-1: Papilloma seen on palate as a white projection (Courtesy Dr Parate).

Diagnosis

- *Clinical diagnosis*—white cauliflower-like projection in oral cavity favors the diagnosis of papillomas.
- *Laboratory diagnosis*—histopathologically it consists of many long, thin, finger-like projections extending from the epithelium.

Differential Diagnosis

- *Verruca vulgaris*—common in skin and always has a sessile base.
- *Papillary squamous cell carcinoma*—base is not pedunculated. It grows rapidly and exceeds 0.5 to 2 cm in size.
- *Verrucous carcinoma*—seen at the age of 60 and 70 years and is associated with smoking and chewing tobacco or snuff, the mass is wider than the surface area with the base almost as wide as the lesion. It does not achieve much vertical length.
- *Condyloma latum and acuminatum*—they are less common and patient has a history of oral sex.

- *Pseudoepitheliomatous hyperplasia*—it has got sessile base.
- *Verruciform xanthoma*—rare in the oral cavity, has a broad base and vertical projections are minimum.

Management

- *Surgical excision*—elliptic incision on the tissue underlying the lesion should be given. Incision should be from the base of mucosa, into which the pedicle or stalk is inserted. If the tumor is properly excised, recurrence is rare.
- *Other modalities*—application of formaldehyde at night on the wart may cure the condition. Sometimes, silver nitrate application also cures the condition.

Verruciform Xanthoma

It is also called as 'Histiocytosis Y'. It is a papillomatous lesion of oral cavity in which accumulation of lipid laden histiocytes occur below the epithelium. The lesion occurs as an unusual reaction to localized epithelial trauma or damage.

Clinical Features

- *Age and sex*—there is strong female predilection and is usually seen in middle age.
- *Site*—it can occur at any site and is most frequently found on the gingiva or alveolar ridge, followed by the buccal mucosa, palate, floor of the mouth, lip and lower mucobuccal fold.
- *Verruciform appearance*—the lesion appears as soft, well demarcated slightly elevated mass with white, yellowish white or red in color. It has got papillary or roughened appearance which is called as verruciform appearance.
- *Crateriform surface*—in some cases, crateriform surface have also been reported.
- *Base*—it is either sessile or has a pedunculated base.
- *Size*—it may be as small as 2 mm to as large as 1.5 cm.

Diagnosis

- *Clinical diagnosis*—the lesion has got verruciform surface and it is usually solitary.
- *Laboratory diagnosis*—biopsy will show large swollen foam cells or *xanthoma* cells, which are presumably histiocytes.

Management

- *Surgical excision*—it is treated with conservative surgical excision.

Keratoacanthoma

It is also called as 'self healing carcinoma', 'molluscum sebaceum', and 'pseudocarcinoma'. It, clinically and

histologically, resembles epidermoid carcinoma and it is frequently mistaken as cancer. It is believed to arise from hair follicles.

Etiology

- *Genetic*—there seems to be hereditary predisposition for the multiple lesions.
- *Viral*—human papillomavirus possible subtypes HPV 26 or HPV 37 have been associated with Keratoacanthoma.
- *Sun exposure*—sun exposure is also one of the etiological factors as it is more commonly seen on sun exposed skin.
- *Chemical*—chemical agents such as coal tar and mineral oil has been associated with Keratoacanthoma in some cases.
- *Trauma*—association of trauma with Keratoacanthoma has been controversial.

Types

- *Ferguson Smith type*—there are large number of keratoacanthoma which is hereditary in nature.
- *Eruptive Grazybowski type*—in this, there are hundreds of small papules on skin and upper digestive tract.

Clinical Features

- *Age and sex*—male to female ratio is 2:1 and majority of cases occur between the ages of 50 to 70 years.
- *Common site*—exposed skin including cheeks, nose and dorsum of the hands. Intraoral lesion is uncommon; if found, is more common on lips.
- *Symptoms*—the lesion is often painful and regional lymphadenopathy may be present.
- *Appearance*—the lesion appears as an elevated umbilicated or crateriform with depressed central core which represent a plug of keratin. Lesion appear to be fixed to surrounding tissue.
- *Shape*—it appears as dome shaped.
- *Color of keratin plug*—keratin pit is frequently discolored, being yellowish brown in color.
- *Lip*—on the lower lip, the lesion shows smooth, raised, rolled borders with a central plug of hard keratin.
- *Margins*—margins are sharply delineated. There may be elevation of the rolled margins.
- *Size*—it grows to maximum size of 1 to 2 cm in diameter.
- *Progress*—it begins as small, firm nodules that develop to full size over a period of four to eight weeks and persist as static lesions for another 4 to 8 weeks. After that, it undergoes spontaneous regression over the next six to eight weeks period by expulsion of the keratin core with resorption of the mass. There may be unsightly scar formation.

- *Muir-Torre syndrome*—it consists of sebaceous neoplasms, keratoacanthoma and gastrointestinal carcinomas.

Diagnosis

- *Clinical diagnosis*—elevated lesion with central keratin plug on sun exposed skin will aid in diagnosis of keratoacanthoma.
- *Laboratory diagnosis*—on biopsy there is thickened layer of parakeratin or orthokeratin with central plugging.

Differential Diagnosis

- *Keratinizing squamous cell carcinoma*—cancerous lesion usually fails to exhibit a smooth round regularity, which is present in keratoacanthoma.
- *Warty dyskeratoma*—is usually small, i.e. less than 0.5 cm, as compared to keratoacanthoma which can attain a dimension of 1-2 cm.

Management

- *Surgical excision*—it often resolves spontaneously without treatment. The lesion may be treated by surgical excision as the scar remaining from excision will be more cosmetic than that resulting from spontaneous regression.

Benign Melanocytic Nevus

It is also called as 'pigmented mole'. Pigmented nevus is a superficial lesion composed of so called nevus cells; hence the term 'cellular nevus'. Nevus is defined as a congenital, developmental tumor like malformation of the skin or mucous membrane.

Types

- *Congenital melanocytic nevus*—These are nevus which are present at birth. Around 15% of congenital nevi is found in head and neck region.
- *Acquired melanocytic nevus (common mole)*—it is present later in life
 - Intradermal nevus
 - Junctional nevus
 - Compound nevus
- *Halo nevus*—it results from nevus cell destruction by immune system.
- *Blue nevus (Jadassohn-Tieche nevus, dermal melanocytoma)*—benign proliferation of dermal melanocytes deep in subepithelial connective tissue.
- *Spitz nevus (benign juvenile melanoma, spindle and epithelioid cell nevus)*—it shares histological features with melanoma. This lesion was described by Spitz in 1948.

Clinical Features

Congenital melanocytic nevus

- *Site*—it is more commonly seen in head and neck region. Intraoral involvement is rare congenital form.
- *Small congenital nevi*—they are greater than 1 cm in diameter and less than 20 cm in diameter.
- *Large nevi*—they are greater than 20 cm in diameter and can cover larger areas of the skin.
- *Appearance*—lesion appear as brown to black plaque with rough surface (**Fig. 15-2**). Early lesion is flat and dark tan which later on becomes elevated, nodular and rougher.
- *Hypertrichosis*—there is presence of excess hair within the lesion. These hairs become prominent with the age giving the appearance of giant hairy nevus.
- *Bathing trunk nevus or garment nevus*—a very large congenital nevus is termed as bathing trunk nevus or garment nevus as patient gives appearance of wearing an article of clothing.



Fig. 15-2: Congenital nevi presented as brown plaque with rough surface.

Acquired melanocytic nevus

- *Age and sex distribution*—acquired nevi are extremely common and appear at 8th month of life and increases in number with age, apparently reaching their peak numerically in the late third decade of life.
- *Site*—it is seen occasionally in the oral cavity, but frequently on the skin. Most of the lesions are seen above the waist. Head and neck are the common site of involvement.
- *Junctional nevus*—this is the earliest presentation of nevus. This name is derived from its location as these are situated in basal layer just above junction of epidermis and dermis. They are flat, demarcated, and brown to black macule. They have regular, oval and round outline.



Fig. 15-3: Compound nevus showing brown elevated lesion above the upper lip.

- **Compound nevus (Fig. 15-3)**—these are slightly elevated, soft papule with smooth surface. The lesion is composed of two parts intradermal and junctional nevus. The degree of pigmentation becomes less with lesion appearing brown or tan.
- **Intradermal nevi**—it may be smooth flat lesion or may be elevated above the surface. It may or may not exhibit brown pigmentation and it often shows strands of hair growing from the surface (**Fig. 15-4**). It is firm on palpation and are raised above the surface. Ulceration is uncommon unless nevi are situated in belt or bra strap area.
- **Intraoral nevi**—intraorally nevus can occur at any site but most commonly occur on hard palate, buccal mucosa, lip, and in gingiva. Most nevi present as raised, macular lesions, but some are flat and macular. Intraoral nevi are slow growing and their size is usually less than 1 cm in diameter.

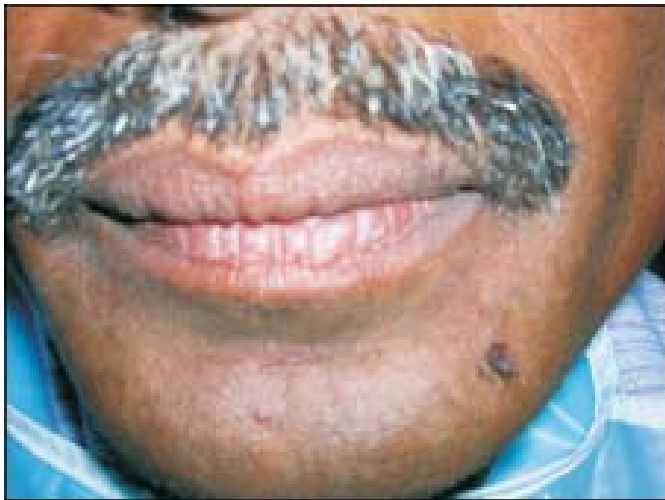


Fig. 15-4: Intradermal nevi showing growing hair from surface.

Halo nevus

- **Site**—it is most commonly seen on skin of trunk.
- **Age**—it is seen in second decade of life.
- **Appearance**—there is central pigmented papule or macule surrounded by zone of hypopigmentation. This zone of hypopigmentation occurs due to destruction of nevus cell by immune system.

Spitz nevus (benign juvenile melanoma, spindle cell or epithelioid cell nevus)

- **Age**—it occurs in children.
- **Site**—it is more commonly seen on the skin of extremities of face.
- **Appearance**—it appears as solitary, dome shaped, pink to reddish brown papule.
- **Size**—it is usually smaller than 6 mm in diameter.

Blue nevus (dermal melanocytoma, Jadassohn-Tieche nevus)

- **Age**—The majority of blue nevi are present at birth and in early childhood. They remain unchanged throughout the life.
- **Site**—it occurs chiefly on buttock, dorsum of the feet and hands, face and occasionally on other areas.
- **Appearance**—The lesions are smooth and exhibit hair growing from the surface.
- **Color**—the color of blue nevi occurs as melanocytes reside deep in the connective tissue and the overlying vessels dampen the brown coloration of melanin and thus yield a blue tint.
- **Intraoral nevi**—in the oral mucosa, they may be macular or nodular in appearance.

Diagnosis

- **Clinical diagnosis**—clinically, nevi can be diagnosed easily by their features.
- **Laboratory diagnosis**—nevus contain granules of melanin pigment in their cytoplasm. Epithelium is thin and irregular and shows “abtropfung” or “dropping off” effect.

Management

- **Surgical removal**—it has been customary to recommend the removal of pigmented mole if it occurs in areas where they are irritated by clothing, such as belt or collar line or if they suddenly begin to increase in size, deepen in color or become ulcerated.

Tumors of Fibrous Connective Tissue Origin

Fibroma

It is a benign soft tissue tumor found in the oral cavity. True benign neoplasm of the fibrous tissue is relatively an

infrequent lesion. Most of these lesions are infact hyperplasia or reactive proliferation of fibrous tissue. It develops due to chronic irritation.

Clinical Features

- *Age and sex distribution*—it can occur at any age but is common in 3rd, 4th and 5th decades. There is no sex predilection.
- *Site*—occurs on the gingiva, tongue, buccal mucosa and palate.
- *Symptoms*—they are usually painless, but if they are in a position where they can be bitten or injured, there may be pain and discomfort.
- *Appearance*—it is most often sessile, dome shaped or slightly pedunculated with smooth contour. The lesions on lips and tongue present as circumscribed nodules.
- *Signs*—tumor sometimes becomes irritated and inflamed and may show superficial ulceration.
- *Surface and color*—the color of tumor is pink and surface is smooth (Fig. 15-5). In some lesion may appear white due to continuous irritation (Fig. 15-6).



Fig. 15-5: Smooth surface pink nodule seen in fibroma of the patient (Courtesy Dr Bande).



Fig. 15-6: Whitish color of fibroma due to chronic irritation.

- *Size*—tumor may be very small or in rare instances may range up to several centimeters in diameter.
- *Consistency*—the consistency can range from soft and myxomatous to firm and elastic. According to the consistency the tumor is termed as 'hard fibroma' and 'soft fibroma'.

Diagnosis

- *Clinical diagnosis*—sessile, firm on palpation with pink color and smooth surface will go in favor of fibroma.
- *Laboratory diagnosis*—biopsy shows bundles of interlacing collagenous fibers interspersed with varying number of fibroblasts or fibrocytes and small blood vessels.

Differential Diagnosis

- *Lipofibroma*—feels softer on palpation.
- *Myxofibroma*—feels softer on palpation.

Management

- *Surgical excisions*—it is treated by conservative surgical excision.

Fibrous Hyperplasia

It is also called as 'inflammatory fibrous hyperplasia', 'denture injury tumor' and 'epulis fissuratum'.

Etiology

- *Ill fitting dentures*—it is the most common reaction to a chronically ill-fitting denture. It occurs due to overextended denture flanges.
- *Others*—other factors which are responsible are ragged margins of teeth, overhanging restorations, sharp spicules of bone, badly fitting clasps and chronic biting of cheek and lips.

Clinical Features

- *Site*—there is development of elongated rolls of tissue in the mucolabial or mucobuccal fold area, into which the denture flanges conveniently fit.
- *Appearance*—there may be small nodular or polypoid overgrowth of fibrous tissue due to gingival irritation.
- *Epulis fissuratum*—when the lesions occur in buccal sulcus due to denture flanges, it is called as epulis fissuratum (Figs 15-7 and 15-8). In it, there is concomitant overgrowth of surrounding fibrous tissues with a groove in it.
- *Signs*—the excess folds of tissue are not usually inflamed clinically, although there may be irritation or even ulceration in the base of the fold, into which the denture flange fits.

- **Consistency**—the lesion is firm on palpation.
- **Size**—the size of the lesion can be small to massive lesion which involves most of the length of the vestibule.
- **Leaf-like denture fibroma or fibroepithelial polyps**—this occurs beneath the denture in hard palate area. It is flattened pink mass with cupped out depression.



Fig. 15-7: Fibrous growth seen in buccal sulcus due to denture in upper tuberosity area (Courtesy Dr Parate).



Fig. 15-8: Fibrous overgrowth seen on the alveolar ridge due to denture.

Diagnosis

- **Clinical diagnosis**—fibrous growth in relation with denture flanges.
- **Laboratory diagnosis**—biopsy shows hyperplastic mass of tissue which is composed of an excessive bulk of fibrous connective tissue covered by a layer of stratified squamous epithelium, may be of normal thickness or show acanthosis.

Differential Diagnosis

- **Hemangioma**—it is present at birth.

- **Metastatic tumor**—there is history of primary tumor.
- **Papilloma**—white with cauliflower-like surface.

Management

- **Elimination of irritation**—elimination of irritation should be done.
- **Excisional biopsy**—it should be treated with excisional biopsy.

Fibrous Epulis

It is the term used when fibrous growth occurs in the gingiva. The possible cause of it is irritation from subgingival calculus or adjacent carious tooth.

Clinical Features

- **Age and sex distribution**—it is more common in females in the ratio of 4:1.
- **Site**—the interdental papilla of the gingiva is the commonest site.
- **Appearance**—the lesion forms a sessile (Fig. 15-9) or pedunculated mass covered by the mucous membrane.
- **Color**—it varies from normal tint to deep red, depending upon the vascularity and inflammatory changes.
- **Signs**—occasionally superficial ulceration can be seen.



Fig. 15-9: Sessile growth seen which arise from interdental papilla in fibrous epulis.

Management

- **Surgical excision**—lesions are excised with small amount of adjacent normal tissue.

Fibrous Histiocytoma

It is also called as 'fibroxanthoma' dermatofibroma, sclerosing hemangioma, and nodular subepithelial fibrosis. It may occur in dermis and rare in oral cavity.

Clinical Features

- *Age and sex*—it is common in young adults, with male predominance.
- *Site*—it is usually seen on dermis (**Fig. 15-10**). Most common site is the extremities where the lesion is called as dermatofibroma. Intraorally it is common on lips, tongue, buccal mucosa and palate.
- *Symptoms*—it is a painless mass.
- *Signs*—there is soft, non-tender, firm swelling of varying size. There is displacement of regional teeth. It is locally an aggressive tumor.



Fig. 15-10: Fibrous histiocytoma showing extraoral swelling.

Diagnosis

- *Clinical diagnosis*—dermal tumor with intraoral soft fibrous growth will go in favor of fibrous histiocytoma.
- *Laboratory diagnosis*—biopsy shows rich vascular growth, made up of histiocytes and collagen producing fibroblast-like cells, which are arranged in a *whorl* or *cartwheel* pattern.

Management

- *Surgical excision*—it can be done and recurrence is uncommon.

Desmoplastic Fibroma

It arises from the mesenchyme of bone. It is also called as 'aggressive fibromatosis'. It produces an abundant number of collagen fibers. It is thought to be counterpart of soft tissue fibromatosis.

Clinical Features

- *Age and sex*—it is most commonly occurs in 2nd decades of life. There is no sex predilection.

- *Site*—in oral cavity, it involves mandible more commonly than maxilla, usually at molar-ramus-angle area. In the skeletal, it involves humerus and tibia.
- *Symptoms*—swelling of the jaw and sometimes, pain and tenderness may be present. In some cases, there may be limited opening of mouth.
- *Signs*—buccal cortex is enlarged and more advanced lesions exhibit facial asymmetry.
- *Progress*—they are aggressive and proliferate rapidly.
- *Syndrome associated*—it may be associated with Gardner's syndrome.

Radiological Features

- *Site*—the most common site is ramus and posterior area of mandible.
- *Radiodensity*—it appears as well defined radiolucency, either unilocular or multilocular. The larger lesions appear to be multilocular with very coarse and thick septa. Smaller lesions are completely radiolucent.
- *Margin*—margin can be well defined or ill defined.
- *Extent*—it may perforate the cortex and infiltrate the adjacent tissues.
- *Teeth*—divergence of contiguous tooth roots is a common finding. In some cases, there may be root resorption.

Diagnosis

- *Clinical diagnosis*—not so specific lesion.
- *Radiological diagnosis*—it should be included in multilocular radiolucent lesion as a differential diagnosis.
- *Laboratory diagnosis*—biopsy shows cells which are small or uniform, plump, or both. The fibrous product is generally mature with thick, wavy collagen fibers arranged in fascicle. The collagen fibers are usually thin and delicate with fasciculation produce a 'herring bone' or 'chevron' or 'storiform' pattern.

Differential Diagnosis

- *Fibrosarcoma*—Coarse and irregular septa and sometimes straight septa are seen in desmoplastic fibroma.

Management

- *Surgical excision*—wide local excision is the treatment of choice in these cases.
- *Curettage*—small lesion is managed by through curettage.
- *Extraction*—involved teeth should be extracted.

Giant Cell Fibroma

It is a well described, benign hyperplastic lesion of oral mucosa. It is not associated with chronic irritation.

Clinical Features

- *Sex and age distribution*—no sex predilection and can occur at any age. It is slightly more prevalent in women.
- *Common sites*—gingiva followed by tongue, palate, buccal mucosa and lips.
- *Symptoms*—it is asymptomatic and may be present for several years.
- *Appearance*—it is usually small, raised, pedunculated, papillary lesion, less than 1 cm in diameter.

Diagnosis

- *Clinical diagnosis*—sessile, pedunculated nodule, less than 1 cm in diameter presence on tongue (**Fig. 15-11**) with no history of irritation will be giant cell fibroma.
- *Laboratory diagnosis*—in biopsy, there is presence of numerous, large stellate and multinucleated giant fibroblasts in the connective tissue, which makes up the bulk of the lesion.



Fig. 15-11: Giant cell fibroma less than 1 cm in diameter seen on tongue.

Management

- *Surgical excision*—surgical excision is done and recurrence is rare.

Myofibroma

This spindle cell tumor consists of myofibroblasts.

Clinical Features

- *Age*—it can occur at any age, but more commonly found in fourth decade of life.
- *Site*—it is more common in head and neck region. Common oral site are lips, cheeks, and tongue.
- *Appearance*—it is painless mass and it exhibits rapid enlargement.

Diagnosis

- *Clinical diagnosis*—not specific.
- *Laboratory diagnosis*—biopsy shows interlacing bundles of spindle cells with tapered or blunt ended nuclei. Tumor cells are positive for smooth muscle stain.

Management

- *Surgical excision*—it is the treatment of choice.

Oral Focal Mucinosi

It is oral counterpart of cutaneous focal mucinosis. This lesion results from overproduction of hyaluronic acid by fibroblast.

Clinical Features

- *Age and sex distribution*—it is common in young adults with slight female predilection.
- *Site*—gingiva is the most common site followed by hard palate.
- *Appearance*—lesion is sessile, painless and nodular mass of same color of surrounding tissue (**Fig. 15-12**).
- *Surface*—it is smooth and non-ulcerated.
- *Size*—size can vary from few mm to 2 cm in diameter.



Fig. 15-12: Oral focal mucinosis showing nodular mass on gingiva which has got sessile base.

Diagnosis

- *Clinical diagnosis*—sessile painless mass on gingiva with same color may suspect this tumor.
- *Laboratory diagnosis*—biopsy shows non-encapsulated area of loose myxomatous connective tissue surrounded by collagenous connective tissue.

Management

- *Surgical excision*—it is the treatment of choice in oral focal mucinosis.

Myxofibroma

Some areas of fibroma undergo myxomatous degeneration.

Clinical Features

- *Site*—it occurs anywhere in the oral cavity, most commonly on palate, lip and gingiva.
- *Signs*—it feels softer than fibroma and is less pale.

Diagnosis

- *Clinical diagnosis*—not so specific.
- *Laboratory diagnosis*—there are regions of dense fibroblastic tissue which are interspersed with pale myxomatous appearing tissue.

Differential Diagnosis

- *Plexiform neurofibroma*—it is spongy and fluctuant.

Management

- *Complete excision*—complete excision of the lesion should be carried out.

Myxoma

Myxoma appears to be true neoplasm which consists of tissue that resembles mesenchymal tissue. Soft tissue myxoma is very rare in the oral cavity.

Clinical Features

- *Age and sex*—it can occur at any age and there is no definite sex predilection.
- *Site*—it occurs on skin of the subcutaneous tissues, genitourinary tract and gastrointestinal tract, or liver, spleen, or even in parotid gland. Intraorally, it occurs on the tongue, buccal mucosa and retromolar area.
- *Appearance*—it is present as soft tissue growth in oral cavity.
- *Nerve sheath myxoma*—it arises from perineural cells of peripheral nerves and contain prominent mucoid matrix. It may be present on tongue, buccal mucosa and retromolar area.

Diagnosis

- *Clinical diagnosis*—it is not specific.
- *Laboratory diagnosis*—biopsy shows loose textured tissue which contains delicate reticulin fibers and mucoid material, probably hyaluronic acid.

Management

- *Surgical*—it is essentially surgical and recurrence is common.

Tumors of Cartilage Tissue Origin

Chondroma

It is a benign cartilaginous tumor. In spite of the fact that mandible and maxilla are membranous bones, they sometimes contain vestigial rests of cartilage.

Types

- *Enchondroma or central*—it develops deep into the bone. It is most commonly seen.
- *Ecchondroma*—it develops on the surface.

Origin

- *Meckel's cartilage*—it is present in the mandibular arch, prior to the appearance of the bone. It usually disappears with the occurrence of ossification in the mandibular arch, but it is possible that the remnants still persist.
- *Fibrocartilage*—in some cases, secondary cartilage like fibrocartilage of mandibular symphysis may persist in the jaw bone and thus can give rise to chondroma.
- *Chondrocranium*—the maxilla develops in close association with chondrocranium. The maxillary sinus develops as an outgrowth from the lateral walls of the nasal capsule. As it grows into the maxilla, it may take remnants of the cartilage from the capsule.
- *Remnants of paraseptal cartilage*—in some cases, remnants of the paraseptal cartilage might persist within the maxilla.

Clinical Features

- *Age and sex*—it occurs in the 5th and 6th decades and males are slightly favored.
- *Site*—it usually occurs in the phalanges and metacarpals. Intraorally, maxilla is slightly favored and it occurs in the anterior region, while in mandible it occurs in premolar-molar region and at symphysis; it may be found in condyle and coronoid process.
- *Symptoms*—it is painless, slowly growing and is locally invasive. Teeth become loose and may be exfoliated.
- *Signs*—the overlying mucosa is seldom ulcerated.
- *Syndrome associated*—it is associated with *Ollier's syndrome*, in which there are multiple enchondromatosis. It is also seen in *Maffucci syndrome* in which there are soft tissue angiomas.

Radiographic features

- *Radiodensity*—irregular radiolucent areas.
- *Margins*—borders may be well defined or ragged or poorly defined.
- *Mottled or blurry appearance*—it may develop radiopacities in osteolytic areas, which produce **mottled (Fig. 15-13) or blurry appearance**.

- *Effect on surrounding structure*—jaw is sometimes expanded by the tumor or the lesion which may cause destruction of the lateral margins, so that it is uncovered by bone in this area. Resorption of involved teeth occurs.
- *Loculated type*—some of the central chondromas may be loculated but it is more likely due to surface strands of bone than due to actual partitions.
- *Ecchondroma*—it is usually situated in the mandibular notch. The coronoid process is directed forward and upward at a much less steep angle, although its length and width are normal.

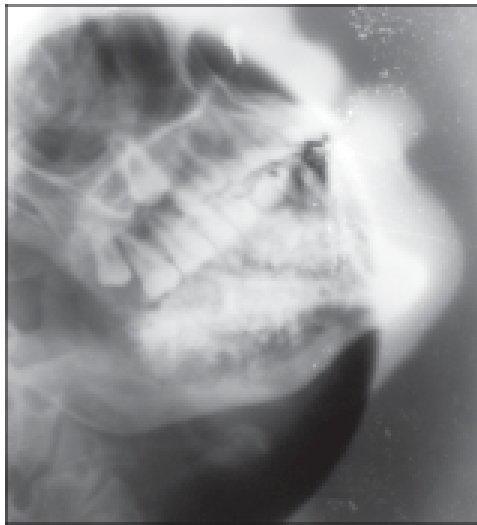


Fig. 15-13: Mottled appearance seen in chondroma in mandibular posterior region (Courtesy Dr Parate).

Diagnosis

- *Clinical diagnosis*—not specific.
- *Radiological diagnosis*—mottled appearance in jaw.
- *Laboratory diagnosis*—biopsy shows hyaline cartilage with calcification or necrosis. Cartilage cells are small.

Differential Diagnosis

- *Chondrosarcoma*—pain is present in case of chondrosarcoma.
- *Osteogenic sarcoma*—typical sun ray appearance is seen.
- *Osteoblastic metastatic carcinoma*—presence of primary tumor.
- *Ossifying sub-periosteal hemangioma*—aspiration should be done to rule out hemangioma.
- *Fibrous dysplasia*—ground glass appearance and histological features.

Management

- *Surgical*—tumor should be excised along with the lining capsule. It should be covered by chip graft. Recurrence is common.

Chondroblastoma

It is also called as 'Codman's' tumor. It usually involves long bone. In some cases, it may be found in cranial bones.

Clinical Features

- *Age and sex*—it occurs in young persons, under the age of 25 years, with male predominance over the female, usually in the ratio of 2:1.
- *Site*—it occurs usually in epiphyseal region of long bones. The usual site is femur and tibia. In some cases, it can occur in condyle of mandible.
- *Symptoms*—it is slow growing, painless mass.

Diagnosis

- *Clinical diagnosis*—not so specific.
- *Laboratory diagnosis*—it consists of uniform, closely packed, polyhedral cells, with occasional foci of chondroid matrix. Multinucleated giant cells are also present.

Management

- *Surgical excision*—conservative surgical excision is carried out. Recurrence is possible after excision.

Chondromyxoid Fibroma

It is an uncommon benign tumor of cartilaginous derivation.

Clinical Features

- *Age and sex*—it occurs in young persons under the age of 25 years, with no definite sex predilection.
- *Site*—it is extremely rare in jaws, but there are some cases reported in mandible. It is more commonly seen in metaphyseal region of long bone.
- *Symptoms*—pain is the outstanding feature of this disease and sometimes, swelling can be seen.

Radiological Features

- *Margin*—margin is sclerotic and scalloped.
- *Appearance*—it is circumscribed radiolucent defect. Central radiopacity is present within the radiolucent defect.
- *Size*—size of lesion varies from 1 to 6 cm.

Diagnosis

- *Clinical diagnosis*—not specific.
- *Radiological features*—radiolucent defect with central radiopacity.

- *Laboratory diagnosis*—biopsy shows myxomatous and fibrous areas and it gives a chondroid appearance. Foci of calcification can be seen.

Management

- *Surgical excision*—conservative surgical excision can be done.

Tumors of Adipose Tissue Origin

Lipoma

It seldom occurs in oral cavity. It is a benign, slow growing tumor composed of mature fat cells. Lipoma appears to be common in obese patient.

Types

- *Encapsulated lipoma*—it is the commonest tumor.
- *Diffuse lipoma*—it does not possess typical features of lipoma. It is also called as 'pseudolipoma'.
- *Lipomatosis*—it has multiple lipomas. It refers to the symmetrical masses of fat, which sometimes occur around the neck in middle aged man and occurs as painful deposits of fat in women in Dercum's disease.

Clinical Features

- *Age and sex distribution*—occurs age after 40 years with peak at 50 years, male to female ratio is 1:1.
- *Common sites*—it usually occurs in upper parts of the trunk, neck (Fig. 15-14) and arms. In oral cavity, it occurs on buccal mucosa and mucobuccal fold followed by tongue, floor of mouth and lip.



Fig. 15-14: Lipoma present on skin in submental region (Courtesy Dr Chole).

- *Appearance*—it appears as a solitary lesion with sessile, pedunculated or submerged base.
- *Size*—size of lesion is approximately 1 cm in diameter.
- *Margins*—well contoured, well defined (Fig. 15-15), round to large ill defined lobulated mass.



Fig. 15-15: Well defined mass of lipoma seen on buccal mucosa (Courtesy Dr Bande).

- *Shape*—it grows as round (Fig. 15-16) or ovoid mass in oral cavity.



Fig. 15-16: Rounded swelling of lipoma seen on buccal mucosa.

- *Base*—it may be lobulated or may be broadly based or have narrow pedicle.
- *Color*—due to thinness of the overlying epithelium, yellow coloration of the fat can be seen.
- *Signs*—surface is smooth, non-tender, soft and cheesy in consistency. The epithelium is usually thin and the superficial blood vessels are readily visible over the surface. Some lesions of lipoma are deep and feel fluid in consistency on palpation; and can be mistaken as cysts.
- *Slip signs*—the edge of lipoma is soft, compressible and often slips away from the examining fingers.
- *Transillumination test*—it may be positive.

- *Lipoblastomatosis*—it is a variant of lipoma, but is not a true neoplasm. It is the continuation of the normal process of fetal fat development carried into the postnatal life. It is characterized by the occurrence in infants. Clinically as a solitary or multiple soft tissue masses, developing at various sites such as the buttocks, chest, axilla or neck.
- *Hibernoma*—it is developed as multi-vacuolated fat that is analogous to the brown fat of hibernating animals; however lesions in oral cavity are not reported.

Diagnosis

- *Clinical diagnosis*—positive slip sign, soft well defined swelling will go in favor of lipoma.
- *Laboratory diagnosis*—biopsy shows mature fat cells, collagen strands coursing through the lesion.

Management

- *Local excision*—surgical excision is done. Recurrence is uncommon.

Bone Tumors

Osteoma

It is a benign neoplasm characterized by proliferation of either compact or cancellous bone, usually in an endosteal or periosteal location.

Origin

- *Periosteal*—it arises on the surface of bone as a pedunculated mass.
- *Endosteal*—it is located in the medullary bone.

Types

- *Ivory osteoma*—it consists of compact bone, which has dense lamellae of bone.
- *Cancellous osteoma*—consisting of trabeculae of bone.
- *Combination*—it is a combination of compact and cancellous variety.

Clinical Features

Ivory osteoma

- *Age and sex distribution*—it occurs in individuals older than 40 years. It is more common in males, as compared to females.
- *Site*—it occurs exclusively on skull and facial bone. It may occur in more than one bone. Mandible is more affected on the lingual side of ramus and inferior border, below the molars.

- *Symptoms*—it is usually painless. Osteoma which arise in sinus can cause sinusitis, headache and ophthalmologic manifestation.
- *Appearance*—asymmetry is caused by bony hard swelling of jaw.
- *Osteoma of condyle*—this may cause deviation of mandible towards the unaffected side (Fig. 15-17).



Fig. 15-17: Osteoma presenting as swelling at angle of mandible (Courtesy Dr Datarkar).

- *Signs*—Mucosa is normal in color and freely movable. Mandibular lesion may be exophytic extending outwards in soft tissues. Ivory osteoma of the jaw is sometimes pedunculated.
- *Osteomatosis*—multiple osteoma may occur as a feature of Gardner's syndrome. Multiple osteoma can also occur in absence of other abnormalities. They have been reported in the mandible, frontal and maxillary sinus. Multiple osteoma rarely occur in soft tissue.

Cancellous osteoma

- *Age and sex*—it more commonly occurs in females with age same as for ivory osteoma.
- *Site*—there is predilection to occur in the alveolar process.
- *Base*—it is usually pedunculated, although it might have a broad base.
- *Surface*—the surface may be smooth or slightly irregular.

Radiographic Features

Ivory osteoma

- *Site*—there is small mass of dense bone situated below the level of the root of lower molar. It may be located within paranasal sinus.
- *Size*—the size of mass is usually few centimeters in diameter.
- *Appearance*—it appears as a uniform radiopaque mass (Fig. 15-18). There may be granular appearance in some

cases. It appears as a homogenous radiodensity without any internal structure.

- *Margins*—it has well defined borders.



Fig. 15-18: Well defined completely radiopaque lesion present due to osteoma.

Cancellous osteoma

- *Internal structure*—it shows evidence of internal trabecular structure. Due to these trabeculations, individual spaces appear to be small. There may or may not be superficial layer of cortical bone.
- *Bone structure*—bone structures, within the tumor, are continuous with that of the parent bone.

Diagnosis

- *Clinical diagnosis*—it is not possible to make clinical diagnosis.
- *Radiological diagnosis*—well defined complete radiopaque lesion will give clue to diagnosis. CT scan may be useful in some cases (**Fig. 15-19**).



Fig. 15-19: CT scan showing osteoma on right side in condylar region.

- *Laboratory diagnosis*—biopsy shows extremely dense compact bone or coarse cancellous bone.

Differential Diagnosis

- *Solid odontome*—there is absence of soft tissue capsule in cases of osteoma and is present in solid odontome.
- *Fibrous dysplasia*—fibrous dysplasia does not reveal the same homogenous density like that of osteoma. Osteomas has usually clearly defined borders.
- *Osteochondroma*—osteochondroma has a cartilaginous capsule and may be associated with irregularity on the surface of the tumor.
- *Sclerosing osteitis*—the margins of sclerosing osteitis tend to be ill defined and in cases of osteoma, margins are well defined. Usually the cause is identified in cases of sclerosing osteitis.
- *Enostosis*—there is absence of any mass on the surface of bone in enostosis.
- *Osteosarcoma*—here, there is bony enlargement with sun-ray appearance.
- *Chondrosarcoma*—same as osteosarcoma.
- *Ossifying fibroma*—bony enlargement with dense radiopaque mass.

Management

- Resection of osteoma is generally successful.

Osteoblastoma

It is also called as 'giant osteoid osteoma'. It is a benign uncommon tumor of osteoblasts, with area of osteoids and calcified tissues.

Clinical Features

- *Age and sex distribution*—male to female ratio is 2:1. Most lesions occur in 2nd and 3rd decades of life.
- *Site*—it more commonly occurs in vertebral column and sacrum. It is rare in jaws and if it occurs, found more commonly in tooth bearing areas of mandible.
- *Symptoms*—it is characterized by pain and swelling of the affected region, which may be of few weeks to a year duration. Pain is not relieved by aspirin as compared to osteoid osteoma.
- *Signs*—there is local expansion of the bone.
- *Aggressive osteoblastoma*—it is characterized by locally aggressive behavior. It is seen in older patient. Pain is present and severe.

Radiographic Features

- *Site*—it is seen in tooth bearing region and temporomandibular joint.

- **Radiodensity**—it may be entirely radiolucent or show varying degree of calcification (**Fig. 15-20**).
- **Margins**—borders may be diffuse or show some signs of cortication. Mandibular lesion may contain a radiolucent halo with outer cortical boundaries.
- **Appearance**—the tumor may show varying degree of calcification which may take a form of sunray or fine granular bone trabeculae.



Fig. 15-20: CT scan of osteoblastoma showing mixed lesion with radiopaque foci in the lesion.

Diagnosis

- **Clinical diagnosis**—bony pain which is not relieved by aspirin with swelling in jaw may suspect osteoblastoma.
- **Radiological diagnosis**—well defined lesion with patchy distribution of mineralization.
- **Laboratory diagnosis**—biopsy shows dilated capillaries, multinucleated giant cells and proliferating osteoblasts.

Differential Diagnosis

- **Osteoid osteoma**—more eccentric radiolucency with sclerotic borders. Histologically, osteoid trabeculae in osteoblastoma are larger, broader and longer, with wider trabecular space than those in osteoid osteoma. An osteoblastoma is more vascular and less painful. Size of osteoblastoma is more than 2 cm and size of osteoid osteoma is less than 2 cm.
- **Osteogenic sarcoma**—benign radiographic appearance of osteoblastoma, as compared to osteogenic sarcoma.

Management

- Curettage and conservative surgical excision. Recurrence may occur.

Osteoid Osteoma

It is a variant of osteoblastoma. It is a true neoplasm of osteoblastic derivative. It is small oval or rounded tumor-like nidus which is composed of osteoid and trabeculae of newly formed bone deposited within the substratum of highly vascularized osteogenic connective tissue tumor. It has usually intracortical lumen. It contains concentration of peripheral nerves which is not seen in other bony lesion.

Clinical Features

- **Age and sex distribution**—it occurs in males between the age of 10 and 15 years.
- **Site**—any part of the skeleton is involved, including the small bone of the hands, feet and vertebrae. The skull and jaws are rarely involved, with slight predilection for the mandible.
- **Symptoms**—the main feature of this neoplasm is severe pain, in spite of the small size of the lesion. Pain aggravates as tumor secretes prostaglandins. Pain usually occurs at night.
- **Signs**—soft tissues over the involved bone area may be swollen and tender.
- **Shape**—it is an oval or round tumor-like lesion.
- **Size**—it has a core of about 1 cm in diameter.
- **Extent**—there tends to be a marked reaction, which may extend for a considerable distance from the tumor itself.

Radiographic Features

- **Site**—in the jaws, it develops in the body of mandible.
- **Appearance**—In some cases, there is a dense area of sclerosis in the bone and it may vary from a narrow zone to a more extensive area of several centimeters.
- **Shape**—small ovoid or round radiolucent area less than 1 cm in diameter.
- **Margins**—these are well defined and surrounded by rim of sclerotic bone.
- **Target-like appearance**—the central radiolucency may exhibit some calcification, which manifests in the form of radiopaque foci on radiographs. This will produce target-like appearance.
- **Effect on surrounding structures**—In occlusal view, the overlying cortex is thickened by new bone being formed subperiosteally.

Diagnosis

- **Clinical diagnosis**—bony pain which is relieved by aspirin with swelling in jaw may suspect osteoid osteoma.
- **Radiological diagnosis**—well defined lesion with size less than 2 cm with target-like appearance will give clue to diagnosis.

- *Laboratory diagnosis*—biopsy shows highly vascularized osteogenic connective tissue. There is central nidus composed of compact osteoid tissue, varying in degree of calcification, interspersed by a vascular connective tissue.

Differential Diagnosis

- *Sclerotic osteitis*—there is presence of central radiolucency, in cases of osteoid osteoma.
- *Ossifying fibroma*—root resorption is rare in osteoid osteoma, but is frequently present in ossifying fibroma. Osteoid osteoma is located more on the inferior aspect.
- *Monostotic fibrous dysplasia*—it is common in maxilla and there is no central radiolucency.
- *Benign cementoblastoma*—the lesion is surrounded by radiolucent halo; while in case of osteoid osteoma, there is central radiolucency.
- *Periapical cemental dysplasia*—it is exclusively found in females.
- *Osteoblastoma*—it is less painful than osteoid osteoma.

Management

- *Salicylates*—severe pain may be relieved by mild obtundants such as aspirin.
- *Excision*—complete excision of tumor is carried out.

Osteochondroma

It is most likely to represent a choristoma, rather than a neoplasm. It is developmental in origin. There is intermingling of two lesions resulting in the term osteochondroma.

Clinical Features

- *Age and sex*—it occurs most often in women, between the ages of 20 and 39 years.
- *Site*—it is commonly seen in lower end of femur, upper end of tibia. It is rare in jaw and if found, it is seen in coronoid process of mandible and mandibular condyle. In some cases, tongue may be involved.
- *Symptoms*—dysphagia may be the only symptom in this patient. If there is involvement of the condyle, there is difficulty in movements of mandible. Pain is experienced, either in opening and closing the mouth or in deviating the mandible to one side.
- *Appearance*—on the tongue, it appears as a pedunculated swelling of about 1 to 2 cm in the posterior part of the dorsum of tongue, near the foramen cecum.
- *Sign*—there is facial asymmetry and lesion has got broad base.

Radiological Features

- *Appearance*—the radiolucent area in the bone tends to be spherical or oval in shape.
- *Margins*—margins of the tumor tend to be more sharply localized and corticated.
- *Internal structure*—it shows the presence of bone in the lesion, unlike chondroma which does not show any bone in the lesion. The bone within the tumor is either trabecular or as irregularly shaped amorphous mass.
- *Coronoid process*—in some cases, coronoid process is enlarged, resembling a *drumstick* appearance.
- *Effect on surrounding structures*—the malar bone may show destruction of bone on its deep and inferior surface, while superficial surface of the adjacent maxilla is indented to receive the additional size of the coronoid.

Diagnosis

- *Clinical diagnosis*—if deviation of mandible during mouth opening, facial asymmetry in the condylar lesion is present, this disease should be suspected.
- *Radiological diagnosis*—drumstick appearance of coronoid process and radiolucent lesion seen in condyle will give clue to diagnosis.
- *Laboratory diagnosis*—biopsy shows a well circumscribed lesion of mature lamellar bone, cartilage or a mixture of those tissues. Haversian canals may be present in the bone and rarely, blood forming elements may be present.

Management

- *Surgical excision*—surgical excision of the lesion should be carried out.

Torus Palatinus

It is also called as '*palatine torus*'. It is a slowly growing flat based bony protuberance or excrescence which occurs in the midline of the hard palate. It occurs in about 20% of the population.

Etiopathogenesis

- *Genetic*—it is inherited as autosomal dominant trait.
- *Functional stress*—environmental factors such as functional stress can also lead to torus palatinus.

Types

They are classified according to their morphology

- *Flat torus*—it has broad base and smooth surface. Extent of this tori is even on both side of midline of palate.
- *Spindle torus*—it has midline ridge along the palatal raphe.

- *Nodular torus*—in this, multiple protuberances with individual base are present. Groove may form in the midline.
- *Lobular torus*—it has lobulated mass with single base. It can be sessile or pedunculated.

Clinical Features

- *Age and sex*—females are affected twice more commonly than males. Development is initiated in young adults, before 30 years.
- *Site*—it occurs in the midline of the palate and may extend to involve the palatal process of the palatine bone.
- *Progress*—the growth may be slow or there may be a period of rather rapid growth which ceases altogether before the patient has advanced into adult life.
- *Size*—they are usually less than 2 cm in diameter. In some cases, lesion may progress to such an extent that it can fill whole palatal surface.
- *Signs*—it is covered with normal mucosa, which appears pale and occasionally ulcerated, when traumatized.

Radiographic Features

- *Radiodensity*—there is relatively dense radiopaque shadow.
- *Site*—if the torus has developed in the middle or anterior regions of the palate, it will be superimposed with apical area of maxillary teeth (**Fig. 15-21**). If it occurs in posterior area, it will appear on the roots of maxillary molars.
- *Margins*—borders are well defined as the surface of the torus is of compact bone.
- *Shape*—there is more or less rounded and sharply defined eminence, which stands well above the palate. Occlusal film that it includes the posterior borders of hard palate will provide good demonstration of palatal torus, which will appear oval in shape.

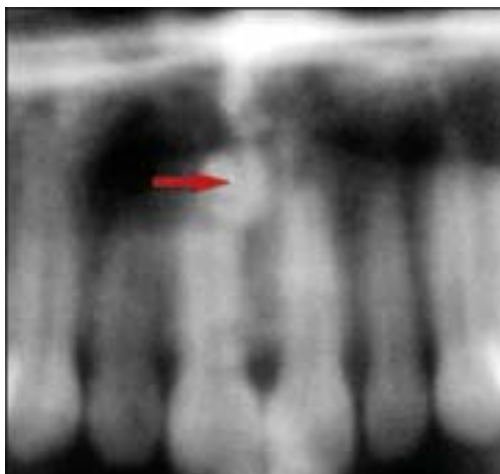


Fig. 15-21: Increased radiopacities seen at the apex of maxillary anterior of palatal tori.

Management

- *Surgery*—as it is benign it is usually not treated, except in some cases where patient requires a complete denture and tori is causing more undercuts and problems for fitting of complete denture. In these cases surgical removal of tori can be done.

Torus Mandibularis

It is also called as '*mandibular torus*'. It is an exostosis or outgrowth of bone found on the lingual surface of the mandible. They consist primarily of compact bone. it occurs in 8% of the population.

Causes

- *Genetic*—it occurs as an autosomal dominant trait.
- *Masticatory stress*—masticatory stress is reported as an essential factor underlying the formation. The reason behind is that torus mandibularis is seen in patient who is having bruxism.

Clinical Features

- *Age and sex distribution*—it is usually discovered in middle aged adults. There is no sex predilection.
- *Site*—it may occur singly, multiply, unilaterally, but is usually bilateral in premolar region.
- *Sign*—there is growth on the lingual surface of the mandible, above the mylohyoid line, usually opposite to the bicuspid teeth (**Fig. 15-22**). Overlying mucosa may get ulcerated in some cases.
- *Size*—their size is variable ranging from an outgrowth that is just palpable to one that contacts a torus on the opposite side.



Fig. 15-22: Torus mandibularis presented as swelling in the bicuspid region (Dr Bande).

Radiographic Features

- **Site**—radiopaque shadow superimposed over the roots of premolars and molars and occasionally, on the incisors and canine.
- **Margins**—they are sharply demarcated anteriorly, on the periapical film and less dense and less well defined as they extend posteriorly.
- **Shape**—the shadow tends to be oval with long axis in the posteroanterior direction.
- **Appearance**—on occlusal radiograph, tori appear as radiopaque, homogenous, knobby protuberances from the lingual surface of mandible. Borders are not sharp between it and bone, but it is continuous suggesting that it is a part of the bone rather than a growth.

Management

- **Surgery**—removal may be necessary, if a mandibular denture is planned.

Exostosis

It is also called as '*hyperostoses*'. They are small regions of osseous hyperplasia of cortical bone and occasionally, cancellous bone, on the surface of the alveolar process. They are less common than mandibular and palatal tori.

Clinical Features

- **Age and sex distribution**—it is more commonly seen in adults as compared young.
- **Site**—it may develop on the palatal surface of maxillary alveolar process, at the border between the attached gingiva and vestibular mucosa, in canine or molar area.
- **Size**—they seldom attain large size and may be solitary or multiple.
- **Shape**—their shape may be nodular, pedunculated or flat protuberance on the surface of bone.
- **Consistency**—they are bony hard on palpation.
- **Buccal exostoses**—it usually occurs as bilateral row of bony hard nodule on buccal aspect of an alveolar ridge. Ulceration of overlying mucosa may occur if trauma occurs.
- **Palatal exostoses (palatal tubercle)**—it is also bilateral and seen in maxillary tuberosity area.
- **Solitary exostoses**—it occurs as reaction of local irritation. Mostly occur below the free gingival graft (**Fig. 15-23**).
- **Reactive subpontine exostosis (subpontic osseous proliferation, subpontic osseous hyperplasia)**—it occurs from alveolar crestal bone beneath the pontic of posterior bridge.

Radiographic Feature

- **Radiodensity**—the internal aspect of an exostosis usually is homogenous and radiopaque.



Fig. 15-23: Solitary exostosis seen on buccal aspect of maxillary molar (Courtesy Dr Abhishek Soni).

- **Appearance**—they appear as circumscribed, smoothly contoured, rounded radiopaque masses.
- **Margins**—they are well defined, but some lesions may poorly blend, radiographically into the surrounding bone.

Management

- **Surgical**—surgical removal is required if exostosis is exposed to frequent trauma and if you want to place prosthesis in that area. Reactive subpontine exostoses need to be removed as they interfere with oral hygiene.

Enostosis

It is also called as '*dense bone island*'. They are internal counterparts of exostosis. They are localized growth of compact bone that extends from the inner surface of cortical bone into the cancellous bone. It is also called as '*whorl*'. A rare condition in which there are thousands of dense islands of bone scattered through the skeleton is known as '*osteopoikile*' or '*osteopoikilosis*'.

Clinical Features

- **Age**—it is more commonly seen in 2nd and 3rd decade.
- **Site**—it is more common in mandible than maxilla and commonly found in premolar molar area.
- **Symptoms**—it is asymptomatic.

Radiographic Features

- **Margins**—single isolated radiopacities that may be either well defined (**Fig. 15-24**) or diffuse, so that the trabeculae blend with trabecular pattern of adjacent normal bone of jaw.
- **Size**—it is more or less rounded, with size varying from a few millimeters up to a centimeter or more.



Fig. 15-24: Single isolated radiopacity seen at the apex of first molar suggestive of enostosis (Courtesy Dr Parate).

- **Teeth**—in rare cases there may be external root resorption of the teeth when it is located periapical to the tooth.

Diagnosis

- **Clinical diagnosis**—not possible to make clinical diagnosis.
- **Radiological diagnosis**—rounded opacity with lack of expansion will give clue to diagnosis.

Differential Diagnosis

- **Periapical idiopathic osteosclerosis**—difficult to distinguish, but these periapical lesions may be associated with periodontal ligament widening, which is absent in case of enostosis.
- **Periapical cemental dysplasia**—radiolucent margins.
- **Hypercementosis**—associated with roots of teeth.
- **Cementoblastoma**—it is also associated with roots of teeth.
- **Osteosarcoma**—radiolucent component is present.
- **Metastatic osteoblastic carcinoma**—radiolucent component is present.
- **Chondrosarcoma**—radiolucent component is present.

Management

- **No treatment**—most cases are unrecognized and are not clinically important.

Tumors of Vascular Tissue Origin

Hemangioma

It is also called as '*vascular nevus*'. It is a benign tumor which occurs most commonly in vertebrae and skull. It is characterized by proliferation of blood vessels. It is often congenital in origin. It is composed of seemingly disorganized vessels that are filled with blood and is connected to the main vascular system.

Types

- **Central**—it occurs in the bone.
- **Capillary hemangioma**—it is a mass of intercommunicating capillary vessels of more or less normal size and structure.
- Strawberry angioma.
- Port-wine stain.
- Salmon's patch.
- **Cavernous hemangioma**—it consists of dilated blood containing spaces, lined by endothelium.
- **Arterial or plexiform hemangioma**—it arises from arteries.

Clinical Features

Central

- **Age and sex distribution**—most of cases are found at birth or arise at early age. Female to male ratio is 2:1.
- **Origin**—the lesion can originate either from the periosteum and resorbs the underlying bone or it occurs within the bone as an anomaly of blood vessels in the marrow spaces.
- **Site**—mandible to maxilla ratio is 2:1. 50% cases are found in mandible, mostly in the body and ramus area.
- **Symptoms**—pain is present in many cases of central hemangioma. The nature of the pain is throbbing. Patient also complaint of swelling in the jaw (**Fig. 15-25**). In some cases, there is paresthesia of skin supplied by mental nerve.



Fig. 15-25: Central hemangioma presented as swelling seen in mandibular posterior region which is red in color (Courtesy Dr Bhaskar).

- **Signs**—swelling is non-tender, compressible and in some cases bony hard in consistency (**Fig. 15-26**). On auscultation, it is pulsatile and bruit may be detected. On probing, there is presence of bleeding from gingiva around the neck of affected teeth. Aspiration of the lesion produces blood.



Fig. 15-26: Central hemangioma presented as hard swelling in the palate.

- *Pumping tooth syndrome*—it demonstrates pumping action, i.e. if the tooth is depressed into the socket, it will rebound into its original position within few minutes.
- *Effect on teeth*—teeth in affected area may be loosened and may migrate.
- *Kasabach-Merritt syndrome*—it is associated with extensive hemangioma in infants. There is also thrombocytopenia and hemorrhage due to platelet trapping within the tumor.

Cavernous

- *Age and sex*—it is common in 1st and 3rd decades and female to male ratio is 2:1. It may fluctuate in size in pregnancy and menarche. The oral or pharyngeal hemangiomas are diagnosed at an older age, than the lesions from other sites.
- *Site*—the most common site of occurrence are the lips, tongue, buccal mucosa and palate.
- *Appearance*—it appears as a flat or raised lesion of mucosa (**Fig. 15-27**).
- *Color*—it is usually bluish in color.
- *Size*—size of swelling vary, and may become larger on physical activity or standing. Size of the swelling reduces in size once the patient is flat on the examining table. Sometimes they may increase in size to such an extent that it can bury the teeth and cause serious deformity and disfigurement.
- *Signs*—Swelling is deep red or bluish red in color and seldom is well circumscribed. In some cases, the texture of mucosa may be more or less unchanged, showing an increased vascularity on the surface; but in some cases, appearance in pebbly.
- *On palpation*—the mucosal hemangioma is typically a soft, moderately well circumscribed lesion. The large lesions are warm and may even be pulsatile if associated with large vessels. The more superficial ones are often lobulated and will blanch under finger pressure. Deeper lesions tend to be dome-shaped with normal or blue surface coloration, they seldom blanch.



Fig. 15-27: Cavernous hemangioma of lip presenting as raised lesion. It is deep red in color.

- *Bleeding upon trauma*—the tumor more often is traumatized and bleeds profusely. It also undergoes ulceration with secondary infection.
- *Base*—some lesions are pedunculated and globular and some are broad based and flat or slightly raised.
- *Compressibility test*—this test is positive. After giving continued pressure and squeezing, blood will drive out of the lesion and the swelling crumbles. As soon as the pressure removed, the swelling reappears with refilling.



Fig. 15-28: Congenital hemangioma presented on tongue which is bluish in color.

- *Tongue*—in cases of hemangioma affecting the tongue, there may be loss of mobility of the tongue. Color of swelling is usually bluish (**Fig. 15-28**). It may affect a part of tongue or the entire tongue.

Capillary Hemangioma

Strawberry angioma

- *Age*—typical history of red mark is noticed after 1-3 weeks of birth. After 1st birthday, it regresses in size and involution is completed by 7 to 8 years.

- *Site*—subcutaneous tissues as well as skin are often involved. In some cases, it can be seen in the floor of mouth.
- *Strawberry appearance*—red mark increases in size for a few months, till it takes a typical strawberry or raspberry-like swelling (**Fig. 15-29**).



Fig. 15-29: Strawberry hemangioma presented as typical strawberry type lesion in the floor of mouth.

- *Color and base*—it is dark red in color. It slightly protrudes from the skin surface as a sessile hemisphere.
- *Compressibility test*—swelling is compressible.
- *Surface*—it is irregular and there may be small areas of ulceration with scabs (**Fig. 15-30**).



Fig. 15-30: Hemangioma of lip showing some areas of ulceration with scabbing.

Port-wine stain

- *Age*—it generally starts at birth and darkens as the child grows, but it does not really grow.
- *Site*—it is common on the face (**Fig. 15-31**) and at the shoulders, neck and buttock.
- *Appearance*—reddish macular hemangiomas are called as port-wine stain. The port wine stain is generally smooth, but could be slightly raised (**Fig. 15-32**).
- *Size*—it is seldom more than 5 mm in diameter.
- *Color*—it is deep purple-red in color, which may become paler later. Color blanches readily on pressure.

Salmon patch

- *Age*—it is present since birth and usually disappears before the first birthday.



Fig. 15-31: Port-wine stain presented as red macular lesion.



Fig. 15-32: Port-wine stain in the distribution of maxillary nerve presented on the face (Courtesy Dr Parate).

- *Site*—it is seen over the forehead, occiput or anywhere in the midline of the body.

Arterial or Plexiform Hemangioma

It is a type of congenital arteriovenous fistula. There is a pulsatile swelling of the arteries, veins become tortuous and thick walled; pulsatile feeling like a bag of pulsating earthworms, is elicited.

Radiographic Features

- *Site*—it is more commonly found in mandible. In mandible they are often located within the inferior alveolar canal. It can also occur in the body and ramus of the mandible.

- *Soft tissue hemangioma*—in some cases if radiograph of patient with hemangioma taken with reduced exposure, the shadow of the lesion may be seen on the radiograph (Fig. 15-33).



Fig. 15-33: Hemangioma of lip presented as increased radiopacity seen on the lower lip (Courtesy Dr Tapasya).

- *Margins*—it is well defined and corticated and in some cases, it may be ill defined.
- *Shape*—the total lesion tends to assume a rounded shape (Fig. 15-34). The bone may show fusiform enlargement at the site of the lesion.

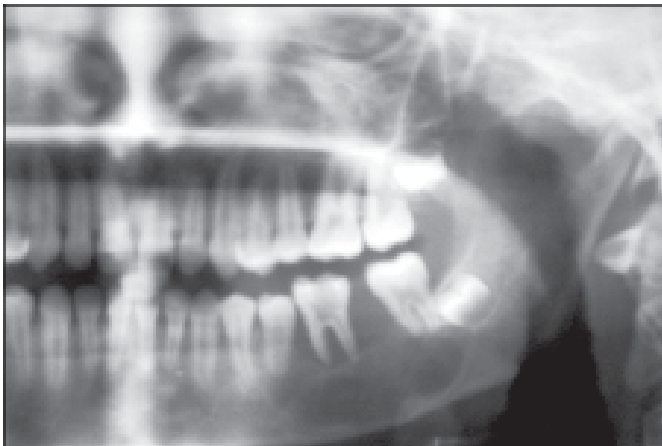


Fig. 15-34: Central hemangioma presenting as rounded lesion in the mandibular posterior region (Courtesy Dr Bhaskar Patle).

- *Soap bubble or honeycomb appearance*—a moderately well defined zone of radiolucency is present within which the trabecular spaces get enlarged and the trabeculae themselves become coarse and thick. The lesion, therefore, presents typically a multicystic, soap bubble or honeycomb appearance (Fig. 15-35).

- *Sunray or sunburst appearance*—the large lesion may cause cortical expansion with radiating spicules at the expanding periphery, occasionally producing the sunray or sunburst appearance.

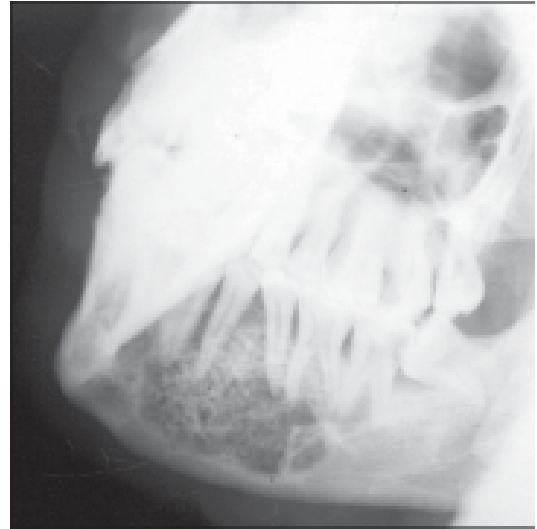


Fig. 15-35: Honeycomb appearance of hemangioma seen in mandibular posterior region.

- *Spokes of wheels appearance*—a structure of the bone is changed in the affected area so that the trabeculae are arranged in a manner which has a rough resemblance to the 'spokes of wheels'.
- *Phleboliths*—small area of calcifications or concretions occasionally within the surrounding soft tissues.
- *Effect on adjacent structures*—roots of teeth adjacent to the lesion are frequently resorbed. Inferior alveolar canal can be enlarged along its entire length and the shape may be changed to a serpiginous pattern. In some cases, the mandibular and mental foramen may be enlarged.
- *Diffuse type of hemangioma*—in it, there is bone destruction giving the appearance of clusters of cavities with ill defined or better defined margins, separated from other of the spaces by normal or rarefied bone.
- *Ultrasound*—ultrasound heterogeneous hypoechoic lesions in which calcified phleboliths are identified, are evident.
- *Computed tomography*—computed tomography shows similar appearance with enhanced quality.
- *CT angiography*—this will determine the extent of blood supply and drainage of the lesion.

Differential Diagnosis (Radiological)

- *Central giant cell granuloma*—central giant cell granuloma crosses the midline, while in hemangioma it does not cross the midline.
- *Giant cell lesions of hyperparathyroidism*—biochemical investigations should be done to rule out hyperparathyroidism.

- *Aneurysmal bone cyst*—hemangioma will show profuse hemorrhage, if aspirated.
- *Ameloblastic fibroma*—in hemangioma, honeycomb pattern and local gingival bleeding with pumping action of tooth are seen.
- *Odontogenic myxoma*—tennis racket pattern is seen in myxoma.
- *Ameloblastoma*—it usually occurs in elder age group.
- *Metastatic tumor*—history of primary tumor is present.
- *Cherubism*—it is usually bilateral and child has a typical facial appearance.
- *Traumatic bone cyst*—it is a well defined entity with better defined borders.
- *Odontogenic keratocyst*—same as traumatic bone cyst.

Management

It usually regresses by itself during adolescent period.

- *Sclerosing technique*—intralesional injections of sclerosing chemicals, such as 1 ml of 5% sodium morrhuate are effective. These agents will produce localized inflammatory reaction with resultant thrombosis, subsequent fibrosis of the endothelial spaces and regression of the lesion. After first injection second injection should be repeated after two weeks. Another agent which is used is 95% ethanol.
- *Surgery*—en bloc resection with ligation of external carotid artery. Sometimes, rash surgical excision of such central lesion of bone results in severe loss of blood, which occasionally leads to exsanguinations of the patient to the point of death. Laser surgery, cryosurgery by dry ice can also be effective.
- *Other injections*—injection of boiling water or hypertonic saline may also be given.
- *Radiation therapy* in the form of external radiation or radium can be used to successfully sclerose the lesion. But risk of inducing neoplastic changes later in life is very high and its use is contraindicated particularly in children.
- *Corticosteroids*—intralesional injections of corticosteroid can be given. This may reduce the size of tumor.
- *Flash lamp pulsed dye laser*—this is useful in case of port wine stain.
- *Embolization*—introduction of inert material into the lesion by vascular route can cause embolization and subsequent regression of the lesion.

Sturge-Weber Syndrome

It is also called as 'encephalotrigeminal angiomatosis'. It is congenital disorder belongs to grouped anomalies collectively known as phakomatoses (mother spot disease).

Classification

1st

- *Complete trisymptomatic*—in this, all three organ systems (eye, skin and CNS) are involved.
- *Incomplete bisymptomatic*—in this, involvement is either oculocutaneous or neurocutaneous.
- *Incomplete monosymptomatic*—in this, there is only neural or cutaneous involvement.

2nd—the Roach scale classification

- *Type I*—both facial and leptomeningeal angioma with glaucoma.
- *Type II*—facial angioma alone without CNS involvement. Some cases having glaucoma.
- *Type III*—isolated leptomeningeal angioma without glaucoma.

Etiology

- *Dysplasia of embryonal vascular system (Royle)*—this will result in hemangiomatosis. It occurs in sixth week of intrauterine life.
- *Developmental insult (Del)*—Sturge-Weber syndrome affects the precursor of tissue which originated in promesencephalic and mesencephalic neural crest. These developmental insults give rise to vascular malformation.
- *Aberrant migration (Crinzi)*—it may arise from aberrant migration of mesodermal and ectodermal elements in the brain and meninges in the fetal growth.

Clinical Features

- *Port-wine stain or nevus flammeus*—it is present at birth. It presents at the distribution of trigeminal nerve. The color varies from light pink to deep purple due to overabundance of capillaries beneath the surface of the involved skin (**Fig. 15-36**).



Fig. 15-36: Port-wine stain seen on left side of face in the patient with Sturge-Weber syndrome (Courtesy Dr Amit Parate).

- **Neurological features**—there is angiomas (excessive blood vessels growth) present at the birth. Patient is present with seizures (twitching of small part of body). Maximum patients of Sturge-Weber syndrome are associated with epilepsy. Prolonged seizures cause neurological injury. In some cases, patient may feel weakness on side opposite to port-wine stain. Headache can also occur.
- **Eyes**—glaucoma (increased pressure within the eye) may be present. Glaucoma occurs due to mechanical obstruction of angle of eyes. If glaucoma is left untreated, it may result in decreased vision and blindness. In some cases, enlargement of the coating of the eye (Buphthalmos) may occur.
- **Oral manifestation**—intraorally it involves oral mucosa. There is presence of vascular hyperplasia of purple color (**Fig. 15-37**). Gingival enlargement can also occur due to vascular hyperplasia.



Fig. 15-37: Intraoral purplish discoloration of oral mucosa on left side in the palate is seen (Courtesy Dr Amit Parate).

Radiological Features

- **Conventional radiography**—it may show classical 'tramlines' or tram-track' or 'trolley-track' calcification in the skull.
- **Computed tomography**—it may show calcification (**Fig. 15-38**). It can also demonstrate brain atrophy, ipsilateral choroids plexus enlargement and abnormal draining veins.
- **Angiography**—it demonstrates lack of superficial cortical veins, nonfilling of dural sinues and abnormal tortus vessels.
- **Other modalities**—other imaging modalities like MRI, SPECT, PET can also be useful.

Management

- **Cosmetic correction**—as port-wine stain is unsightly, it should be treated for cosmetic correction. The lesion can be covered by camouflage make up by dermatologist.



Fig. 15-38: Intracranial calcification seen in the brain (arrow) on the computed tomography.

- **LASER**—LASER treatment should be done to lighten and to remove port-wine stain. LASER treatment should start when the lesion is small.
- **Anticonvulsant drug**—it should be given to control seizures.
- **Anti-glaucoma medication**—topical anti-glaucoma therapy for extended period of time is helpful.
- **Dental management**—due to Sturge-Weber syndrome, hemorrhage can occur. So in dental office it should be managed carefully.

Lymphangioma

It is a benign hamartomatous proliferation of lymphatic vascular tissue. It is a hamartoma, rather than a neoplasm. In it, abnormal vessels are filled with clear protein rich fluid containing lymph rather than blood.

Types

- **Superficial or lymphangioma simplex (capillary lymphangioma)**—it presents a circumscribed lesion, which appears as small blisters and slightly elevated skin patches. It involves capillary size vessels.
- **Cavernous lymphangioma**—it is composed of larger dilated lymphatic vessels.
- **Cystic or deep lymphangioma**—they are large, cystic, and translucent and may be seen in the neck, mediastinum or axilla. These are called as *cystic hygroma*.

Clinical Features

- **Age**—they may present at birth with majority becoming clinically evident early in life, but with a small number not being manifested for a number of years.

- **Incidence**—it may occur alone or in association with hemangioma or other anomalous blood vessels.
- **Site**—it occurs in dorsal and lateral borders of tongue, lips, gingiva and buccal mucosa. On face it can involve eyes (**Fig. 15-39**).
- **Symptoms**—usually the disfigurement is noticed by the child parents. Occasionally, the vesicles may be rubbed with clothes, get infected and become painful.
- **Appearance**—they are soft masses that dissect along the tissue planes and turn out to be more extensive than anticipated.
- **Surface**—the surface of the lesion may be smooth or nodular.
- **Color**—color ranging from normal mucosal pink to bluish and may be quite translucent.



Fig. 15-39: Lymphangioma affecting left eye of the patient (Courtesy Dr Bhaskar Patle).

- **Signs**—they are liable to trauma. Due to this, lesions are subjected to periodic attacks of inflammation which cause the swelling to become larger and tender for the time being.
- **Aspiration**—aspiration yields lymph that is high in lipid.
- **Tongue**—if the tongue is affected, enlargement may occur and the term 'macroglossia' is applied. On the tongue, it is characterized by irregular nodularity of the surface of the tongue with gray and pink, grape-like projection. They are often elevated and nodular in appearance and may have the same color as the surrounding mucosa.
- **Lip**—lip involvement and its deformity is called as macrocheilia.

Radiological Features

- **CT feature**—this will show destructive lesion. Contrast CT will show a mass in the lesion (**Fig. 15-40**)



Fig. 15-40: CT scan of lymphangioma of eye showing destructive lesion of left eye (Courtesy Dr Bhaskar Patle).

Diagnosis

- **Clinical features**—irregular nodular lesion of tongue can give clue to diagnose lymphangioma.
- **Laboratory diagnosis**—capillary types are composed of proliferation of thin walled endothelium-lined channels, primarily devoid of erythrocytes. Cavernous type is characterized by presence of dilated sinusoidal endothelium-lined vascular channels, devoid of erythrocytes.

Management

- **Surgical removal**—it is done to remove bulk of the lesion. Partial or complete spontaneous involution is occasionally noted.
- **Sclerosing agent therapy**—as such it does not respond to sclerosing agents. But some success has been reported by using OK-432 a lyophilized incubation mixture of low virulent strain of *Streptococcus pyogenes* with penicillin G potassium which has lost streptolysin S producing ability.

Arteriovenous Fistula

It is also called as 'arteriovenous shunt' or 'arteriovenous malformation'. It is a direct communication between an artery and vein that bypasses the intervening capillary bed. It may be congenital or acquired.

Classification

- **Cirroid aneurysm**—it is a tortuous mass of small arteries and veins linking a larger artery and vein.

- *Varicose aneurysm*—it consists of endothelium-lined sac connecting an artery and a vein.
- *Aneurysmal varix*—it is a direct connection between artery and vein.

Clinical Features

- *Sites*—head and neck are the most common site. It can occur in alveolar ridge, palate and soft tissues.
- *Symptoms*—there is mass of extraosseous soft tissue swelling.
- *Color*—it is having purplish discoloration.
- *Signs*—bone may be expanded. Pulse may be detected on auscultation and aspiration produces blood.
- *Post-extraction bleeding*—it will result in severe bleeding after the extraction of tooth. In some cases it can be life threatening.

Radiographic Features

- *Site*—it develops in the ramus and retromolar areas of the mandible and may also involve the mandibular canal.
- *Radiodensity*—it causes a resorptive radiolucent lesion.
- *Margins*—they are well defined and corticated.
- *Appearance*—it is multilocular. Walls of shunt may contain apparent calcified material.
- *Angiography*—it is seen as abnormal collection of vessels located in the suspected area, with many vessels feeding and draining the lesion.

Diagnosis

- *Clinical diagnosis*—a lesion with a thrill or bruit, or with an obviously warmer surface, is most likely a special vascular malformation, called arteriovenous malformation
- *Radiological diagnosis*—irregular alveolar canal will give clue to the diagnosis. Angiography (**Fig. 15-41**) and contrast study CT scanning will be helpful in conforming the diagnosis.

Differential Diagnosis

- *Hemangioma*—soft tissue hemangioma does not involve the bone.
- *Ameloblastoma*—aspiration of arteriovenous shunt will produce blood which is not seen in tumor.
- *Radicular cyst*—it is associated with a non-vital tooth.
- *Dentigerous cyst*—it surrounds the crown of the tooth.

Management

- *Surgical excision*—it should be treated by surgical excision.

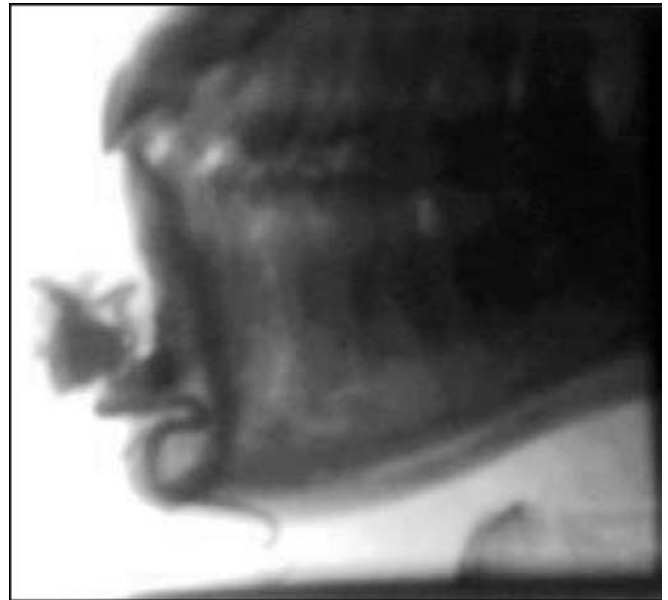


Fig. 15-41: Angiography showing abnormal collection of vessels (Courtesy Dr Datarkar).

Glomus Tumor

It is also called as 'glomangioma'. It is a rare neoplasm derived from glomus cells. They are thought to be closely related to hemangiopericytoma. The glomus is arteriovenous anastomosis that controls the blood supply and temperature of the skin and certain deeper tissues. These functions appear to be mediated in some way by the rich nerve supply and by certain epithelioid cells that ensheath the arteriole of the glomus. The epithelial cells are thought to be comparable to pericytes.

Clinical Features

- *Age*—the tumor usually occurs in the 5th decade.
- *Site*—the tumor probably arises from these specialized glomus cells and occurs most frequently under the nails and on the body surface especially in head and neck area. In the oral cavity the lesion is usually located on the dorsum of the tongue, lip, palate, buccal mucosa and tongue.
- *Size*—they are small lesions, rarely exceeding 1 cm in diameter.
- *Symptoms*—they often give rise to attacks of very severe pain and are exquisitely tender. Pain is stabbing in nature.
- *Color*—the color varies from deep red to purple or blue.

Diagnosis

- *Clinical diagnosis*—severe and stabbing pain with deep red color lesion will give clue to the diagnosis

- *Laboratory diagnosis*—biopsy shows glomus cells which arranged around the blood vessels in a manner suggesting hemangiopericytoma or the blood vessels may be so prominent as to resemble cavernous hemangioma.

Management

- *Surgical excision*—it is a benign tumor and removal of this surgically effects the cure.

Hemangiopericytoma

It is characterized by the proliferation of capillaries surrounded by mass of round or spindle shaped cells which are called as pericytes. It resembles to Glomus tumor but lack its organoid pattern, encapsulation and clinical manifestation of pain.

Clinical Features

- *Age and sex*—there is no sex predilection with age ranging from birth to old age.
- *Site*—it can occur at any site in the oral cavity.
- *Symptoms*—it is usually painless.
- *Appearance*—the lesions are firm, apparently circumscribed and often nodular. It may or may not exhibit redness indicative of vascular nature.
- *Progress*—majority of tumors grow rapidly and are therefore of short duration.

Diagnosis

- *Clinical diagnosis*—not so specific.
- *Radiological diagnosis*—biopsy shows proliferation of occult capillaries. Each vessel in turn is surrounded by a connective tissue sheath, outside of which are found mass of tumor cells. The tumor cells may appear large or small, round or spindle shaped and show tendency for concentric layering about the capillaries.

Management

- *Surgical excision*—local excision of the tumors is treatment of choice.

Nasopharyngeal Angiofibroma

It is also called as *juvenile nasopharyngeal angiofibroma* as its presence in younger age group. Nasopharyngeal angiofibroma is an uncommon, highly vascular, non-encapsulated polypoid mass that is histologically benign but locally aggressive. Hormonal influence is also present in this tumor.

Clinical Features

- *Age and sex*—it is more common in younger age group within the range of 10 to 18 years. It is exclusively found in males.
- *Site*—more commonly affected sphenoid sinus followed by maxillary and ethmoid sinuses.
- *Symptoms*—nasal obstruction, epistaxis, facial deformity (**Fig. 15-42**), proptosis, nasal voice, sinusitis, nasal discharge, serous otitis media, headache and anosmia.
- *Signs*—it is slow growing angiofibroma and growth is asymmetric.



Fig. 15-42: Facial deformity seen in angiofibroma (Courtesy Dr Ashok L).

Radiographic Features

- *Plain radiography features*—there is soft tissue nasopharyngeal mass. There is also widening of the pterygopalatine fossa with anterior bowing of the ipsilateral posterior antral wall. A polypoid nasal mass may cloud the ipsilateral ethmoid and maxillary sinuses.
- *Contrast-enhanced CT*—enhancing mass with appearance same as plain film radiography. Intraorbital and intracranial extension are much better visualized.
- *MRI*—it shows intermediate signal intensity on T₁ weighted and T₂ weighted sequences.

Diagnosis

- *Clinical diagnosis*—features of nasal obstruction can give some clue to this diagnosis.
- *Radiological features*—on CT, enhancing mass will present with intraorbital and intracranial extension.
- *Laboratory diagnosis*—biopsy shows dense fibrous connective tissue which contain numerous dilated thin walled blood vessels of variable size.

Management

- *Surgical excision*—surgical excision is the treatment of choice depending upon extent of lesion. Preoperative embolization will help to prevent blood clot.
- *Radiotherapy*—it is advised for recurrent lesion.

Olfactory Neuroblastoma

It is also called as '*esthesioneuroblastoma*'. It is a neural crest derived neoplasm, arising from the olfactory mucosa in the superior nasal fossa.

Clinical Features

- *Age*—it has got bimodal age distribution with first incidence in 2nd decade and 2nd incidence in 6th decade.
- *Symptoms*—the most common symptoms are nasal obstruction, epistaxis and pain.
- *Appearance*—it appears as solitary mass, sessile or pedunculated, which bleeds profusely.

Kadish Staging System

- Stage A—confined to nasal cavity.
- Stage B—nasal cavity + one or more paranasal sinuses.
- Stage C—extending beyond nasal cavity and paranasal sinuses.

Radiological Features

- *CT*—it appears as homogenous, enhancing mass that primary remodel the bone. It commonly extends into ethmoid and maxillary sinus and rarely into sphenoid sinus. Gross calcifications occur within the tumor mass.
- *MRI*—On MRI, there will be intermediate signal intensity and high T₂ weighted signal intensity are found.

Diagnosis

- *Clinical diagnosis*—not specific.
- *Radiological diagnosis*—on computed tomography homogenous mass in sinus may suspect this tumor.
- *Laboratory diagnosis*—biopsy shows small, round to ovoid basophilic cells which are arranged in sheets and lobules.

Management

- *Surgical excision*—it is the treatment of choice. This surgical excision is also combined with adjunct radiation therapy.

Tumors of Neural Tissue Origin

Neurofibroma or Neurofibromatosis

Many authors described the neurofibroma and neurofibromatosis as different entity. The difference between neurofibroma and neurofibromatosis is that neurofibroma occurs as solitary lesion without café au lait spot and neurofibromatosis occurs as multiple lesions with café au lait spot. Neurofibromatosis is called as '*Von Recklinghausen's disease of skin*' or '*fibroma Molluscum*'. It is inherited as a simple autosomal dominant trait with variable penetrance.

Origin

- *Schwann cells and axon*—it arises from the connective tissue sheath of Schwann cells and axons.
- *Sites of origin*—there are three sites of origin: within the inferior dental canal, in the substance of the bone and beneath the periosteum.

Clinical Features

- *Age and sex distribution*—it occurs at any age but is found more in younger group with no sex predilection.
- *Site*—most commonly affected sites are trunk, face and extremities.
- *Appearance*—it may appear as numerous sessile (**Fig. 15-44**) or pedunculated, elevated smooth surfaced nodules of variable size, which are scattered over the skin surface. In some cases, loose overgrowth of thickened, pigmented skin may hang in folds (**Fig. 15-43**).
- *Elephantiasis neuromatosa*—in other forms, there are deeper, more diffuse lesions which are often of greater proportion than superficial nodules and are sometimes referred to as '*elephantiasis neuromatosa*'.



Fig. 15-43: Overgrowth of thickened pigmented skin seen on face.



Fig. 15-44: Numerous sessile nodules seen on patient body suggestive of neurofibroma (Courtesy Dr Milind Chandurkar).

- *Bags of worms*—neurofibromatosis lesion may feel like bags of worms which is pathognomonic sign.
- *Café au lait spots*—in addition majority of the patients exhibit asymmetric areas of cutaneous pigmentation, often described as “café au lait” spots. The spots are smooth edge, yellowish tan to dark brown macules. They vary in diameter from 1 to 2 mm.
- *Crowe’s signs*—axillary freckling (brown spot on skin) is known as Crowe’s sign.
- *Lisch nodules*—it is translucent brown pigmentation on the iris.
- *Complication*—it can develop into malignant peripheral nerve sheath tumor (Neurofibrosarcoma).

Oral Manifestations

- *Common sites*—it may occur in mandibular canal, buccal mucosa, and alveolar ridge and below the periosteum.
- *Central lesion*—the central lesion may have multiple lesions, occurring in both jaws simultaneously, expanding and filling the maxillary sinus. Solitary central lesion may infrequently be associated with brown spots on skin.
- *Symptoms*—it may produce pain or paresthesia, if associated with mandibular nerve.
- *Appearance*—there are discrete, non-ulcerated nodules, which tend to be of the same color as the normal mucosa (**Fig. 15-45**).
- *Signs*—it may expand and perforate the cortex, producing a swelling that is either hard or firm on palpation.

- *Tongue*—macroglossia may be due to diffuse involvement of the tongue.



Fig. 15-45: Discrete non-ulcerated nodules seen in oral cavity due to neurofibromatosis.

Radiographic Features

- *Extraperiosteal neurofibroma*
 - *Appearance*—there is an area of bone destruction which presents a radiolucent shadow of varying density, depending on the amount of bone that has been replaced by tumor.
 - *Size*—the size of the lesion is variable from less than one centimeter up to a very large lesion.
 - *Margins*—margins of radiolucency are well defined, curved and may be hyperostotic.
- *Subperiosteal neurofibroma*—in some cases the periosteum lays down a layer of bone on the superficial aspect of the tumor. Tumor that causes relatively little indentation of the surface of the bone appears as radiolucent areas, the darkness depending on the depth of bone loss; deeper lesion causing greater darkness in the radiograph than the superficial ones. There is tendency for the shadow to be more or less circular and there may be well defined margins, with or without a bony cortex.
- *Neurofibroma of inferior dental nerve*—it shows fusiform or more or less circular enlargement of mandibular canal. In some cases there may be an elongated lesion with an undulating border.

Diagnosis

- *Clinical diagnosis*—pigmented swelling with café au lait spots.
- *Radiological features*—radiolucent lesion with corticated border (**Fig. 15-46**).
- *Laboratory diagnosis*—it is composed of proliferation of delicate spindle cells with thin wavy nuclei intermingled with neurites in an irregular pattern as well as delicate intertwining connective tissue fibrils.

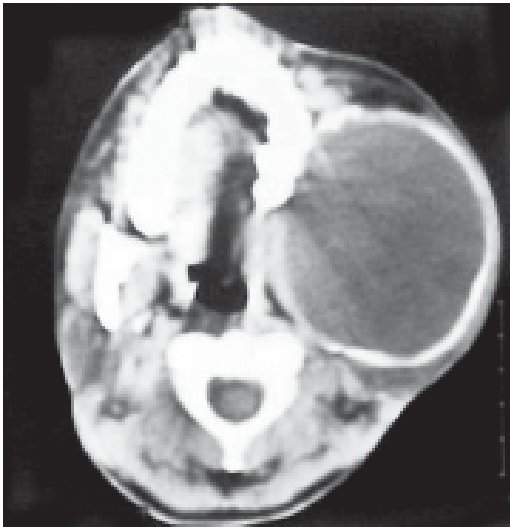


Fig. 15-46: CT scan of neurofibroma showing circular radiolucency with corticated border.

Differential Diagnosis

- *Vascular lesion*—changes in the canal from neural lesion are more localized and have a fusiform enlargement, while the vascular lesion enlarges the whole canal and alters its path.

Management

- *Surgical excision*—solitary lesion may be surgically excised.
- *Extensive lesion*—in this carbon dioxide laser and dermabrasion have been used successfully.
- *Prognosis*—it has got high potential for malignant change.

Schwannoma

It is also called as 'neurilemmoma', 'perineural fibroblastoma', 'neurinoma' and 'lemmoma'. But nowadays all above terms are not used and Schwannoma is the only term used. It is of neuroectodermal origin, arising from Schwann cells that makes the inner layer covering the peripheral nerves.

Clinical Features

- *Age and sex*—occurs at any age, from very young to very old, with equal frequency in both the sexes.
- *Site*—the tumor usually occurs in the subcutaneous tissue, but internal organs such as stomach may be affected. Intraorally, mandible and tongue is the most commonly affected site for central lesion. Other sites which can be involved in these tumors are palate, floor of mouth and buccal mucosa.
- *Progress*—it is a slow growing lesion and is usually of long duration at the time of presentation.

- *Symptoms*—usual complaint is lump in jaw, in case of central tumor and single circumscribed nodule, in case of soft tissue lesions. Paresthesia may be associated, which occurs anterior to the tumor. Pain is localized to the tumor site.
- *Extent*—usually occurs singly and jaw expansion may lead to perforation.
- *Consistency*—the mass is firm on palpation.
- *Aspiration*—it is non-productive to aspiration.



Fig. 15-47: Oval radiolucent area of bone destruction with hyperostotic border in case of neurilemmoma (Courtesy Dr Ashok L).

Radiographic Features

- *Site*—the radiolucent area is usually located within the expanded inferior alveolar nerve, posterior to the mental foramen.
- *Shape*—round to oval radiolucent area of bone destruction (Fig. 15-47). There may be crescent shaped cystic radiolucency.
- *Margins*—margins of the lesions are well defined and hyperostotic.
- *Effect on surrounding structures*—it may cause root resorption of adjacent teeth (Fig. 15-48). If the tumor protrudes from the mental foramina, erosive lesion of the surface of jaw bone occurs. The tumor may expand the inferior alveolar canal, mental foramen, mandibular foramen and mandible while maintaining the cortical boundaries.



Fig. 15-48: Root resorption associated with neurilemmoma (Courtesy Dr K Patil).

Diagnosis

- *Clinical diagnosis*—paresthesia with lump in the mandibular posterior region will suspect neurilemmoma.
- *Radiological features*—oval radiolucent lesion with corticated border.
- *Laboratory diagnosis*—biopsy shows two types of tissues: Antoni type A and Antoni type B.

Differential Diagnosis

- *Cysts*—in neurilemmoma there is an expansion of the inferior alveolar nerve canal.
- *Ameloblastoma*—it usually occurs above the canal.
- *Vascular lesion*—the expansion of the canal caused by neural neoplasm is more concentric, creating a fusiform shape; whereas vascular lesion increases the girth of the canal down the entire length and often alters the shape into a serpiginous form.

Management

- *Surgical excision*—it is the treatment of choice.

Neuroma

It is also called as 'amputation neuroma' or 'traumatic neuroma'. It is not a true neoplasm, but an exuberant attempt at repair of a damaged nerve trunk.

Etiopathogenesis

- *Causes*—nerve damage may result from fracture, dissection, removal of cyst, nerve avulsion for neuralgia or even extraction of teeth.
- *Overgrowth of nerve*—it is an overgrowth of severed nerve, attempting to regenerate when the scar tissue or malalignment of a fractured nutrient canal blocks the distal end.
- *Unorganized collection of nerve fibers*—proliferating nerve forms unorganized collection of nerve fibers, composed of varying proportion of axons, perineural connective tissue and Schwann cells.

Clinical Features

- *Age and sex distribution*—it can occur at any age and it is more common in females.
- *Site*—it typically occurs near the mental foramen, on the alveolar ridge in edentulous areas or on the lips and tongue.
- *Appearance*—it appears as a small nodule or swelling of the mucosa.
- *Symptoms*—due to the pressure applied by enlargement of the tangled mass in its bony cavity, severe pain may be experienced. It may have reflex neuralgia with pain referred to the eye, face and head.

- *Size*—it is a slow growing reactive hyperplasia that seldom becomes large, rarely with an excess of 1 cm in diameter.

Radiographic Features

- *Site*—in mandible it is associated with the mandibular canal. In maxilla, it is seen in anterior maxilla.
- *Radiodensity*—it is seen as a destructive lesion. It appears as a radiolucent area.
- *Margins*—it has got well defined and corticated borders (Fig. 15-49).
- *Inferior alveolar nerve canal*—some expansion of the canal is seen.



Fig. 15-49: Neuroma seen in mandible in inferior alveolar canal region as well defined and corticated margin.

Diagnosis

- *Clinical diagnosis*—severe pain, nodule in mental foramina area with history of extraction will aid in diagnosis of traumatic neuroma.
- *Radiological diagnosis*—radiolucent lesion in the mandibular region with corticated border.
- *Laboratory diagnosis*—biopsy shows mass of irregular and interlacing neurofibrils and Schwann cells, situated in connective tissue stroma of either scanty or plentiful proportion.

Differential Diagnosis (Radiological)

- *Odontogenic cyst*—when an individual has painful or extremely sensitive cyst-like radiolucency in the bone with the history of fracture or surgery, neuroma is suspected.

Management

- *Surgical excision*—simple excision of nodule along with proximal portion of the involved nerve.

Palisaded Encapsulated Neuroma

It is also called as 'solitary circumscribed neuroma'. It is reactive lesion rather than true neoplasm.

Clinical Features

- *Age and sex distribution*—it is more commonly seen in fifth and seventh decade of life with no sex distribution.
- *Site*—nose, cheek and intraorally hard palate is common site.
- *Appearance*—it has smooth surface, painless, dome shaped papule or nodule.
- *Size*—it is less than 1 cm in diameter.

Diagnosis

- *Clinical diagnosis*—not so specific.
- *Laboratory diagnosis*—biopsy shows interlacing fascicles of spindle that consist of Schwann cells. Nuclei are wavy and pointed. Palisading and Verocay bodies of Antoni A cells are present.

Management

- *Surgical excision*—conservative surgical excision should be done.

Ganglioneuroma

It is same like neuroblastoma, but in it, differentiated cells are numerous. It grows less rapidly than the neuroblastoma and when fully differentiated, it depicts benign characteristic. The fully differentiated ganglioneuroma is composed of cells that are very much like the normal cells. Numerous nerve fibers and well differentiated nerve bundles are present.

Melanotic Neuroectodermal Tumor of Infancy

It is also called previously as 'pigmented ameloblastoma', 'melanoameloblastoma', 'melanotic ameloblastoma', 'retinal anlage tumor', and 'melanotic progonoma' due to possible source of tumor from odontogenic epithelium. Nowadays, these terms are not used as it is proved that tumor is of neural crest in origin.

Clinical Features

- *Age and sex*—it occurs in infants under the age of six months, with equal sex distribution.
- *Site*—maxilla is more commonly affected than mandible. It is more common in anterior region.
- *Symptoms*—it is symptomless tumor.
- *Appearance*—it is rapidly growing, non-ulcerated, darkly pigmented lesion. It is blue black in color.

Radiological Features

- *Radiodensity*—there may be well defined areas of radiolucency. Destruction of underlying bone occurs.
- *Margins*—margins of these lesions are irregular.
- *Teeth*—displacement of developing teeth is present.
- *Sunray appearance*—in some cases there may be osteogenic reaction giving sunray appearance.

Diagnosis

- *Clinical features*—blue black mass in anterior maxilla in infant will give clue to diagnosis.
- *Laboratory diagnosis*—high urinary level of vanillyl-mandelic acid (VMA). Biopsy shows both pigmented and non-pigmented cells.

Management

- *Surgical removal*—conservative surgical excision should be carried out. Recurrence is extremely low.

Muscle Tumors

Leiomyoma

It is a benign tumor derived from smooth muscle and is found in a variety of anatomic sites like skin and subcutaneous tissues. It can be solid leiomyoma, vascular leiomyoma and epithelioid leiomyoma.

Clinical Features

- *Age and sex*—it occurs in middle decades of life. Males are affected more commonly than females.
- *Site*—it usually occurs in uterus. It is uncommon in oral cavity due to general absence of smooth muscles except in blood vessel walls and circumvallate papillae of tongue. The smooth muscle of the arrectores pilorum may be a source of cheek tumor.
- *Symptoms*—the patient may complain of sore throat or tumor in the throat. In some cases, there may be pain.
- *Appearance*—it is a slow growing, painless lesion, which is superficial and often pedunculated.
- *Size*—in most of the cases, the lesion is small.
- *Color*—it does not ulcerate and resembles the normal mucosa in color and texture. In case of vascular leiomyomas it can exhibit bluish hue.

Diagnosis

- *Clinical diagnosis*—it has got no typical clinical features.
- *Laboratory diagnosis*—biopsy shows interlacing bundles of smooth muscle fibers, interspersed by varying amounts of fibrous connective tissue.

- *Angioleiomyoma*—in some cases, origin from the blood vessels is obvious since the vessels are enlarged with thick muscular walls, around which the tumor muscle fibers are dispersed in a circular manner. It is called as angioleiomyoma or angiomyoma.
- *Angiomyolipoma*—rarely, angiomyoma may contain adipose tissue: then it is called as angiomyolipoma.

Management

- *Surgical excision*—it is treated by conservative surgical excision of the tumor.

Rhabdomyoma

It is a benign tumor of striated muscle origin. It is an exceedingly rare lesion.

Clinical Features

- *Age*—most commonly occurs in 5th decade of life with male to female ratio of 2:1.
- *Site*—it mostly occurs in head and neck region. The most common sites of occurrence, intraorally, are the floor of the mouth, tongue, soft palate, buccal mucosa and lower lip.
- *Symptoms*—it is painless and slow growing. Laryngeal and pharyngeal lesion may lead to airway obstruction.
- *Appearance*—it presents a well circumscribed tumor mass which may have a known duration of months or even several years.

Diagnosis

- *Clinical diagnosis*—it has got no typical clinical features.
- *Laboratory diagnosis*—the nucleus is vesicular and cells with several nuclei are sometimes seen.
- *Adult's type*—in adult type, the tumor is composed of large, round cells that have granular, eosinophilic cytoplasm and show irregular cross striations. The cytoplasm is rich in glycogen and glycoprotein.
- *Fetal*—fetal type is characterized by immature skeletal muscles in varying stages of development and undifferentiated mesenchymal cells.

Management

- *Surgical excision*—it is excised conservatively usually enucleating with ease.

Granular Cell Tumor

It is also called as '*myoblastic myoma*', '*granular cell myoblastoma*' and '*granular cell schwannoma*'.

Origin

- *Striated muscle*—it may be derived from striated muscles. But, these tumors may be found in areas like breast and skin, where the striated muscles are absent.
- *Neural theory*—in neural theory it is proposed that these tumors are derived from the connective tissue of nerves and hence was called as '*granular cell neural fibroma*'.
- *Stem cells*—some authors state that it is derived from stem cells with a leiomyofibrillogenic capacity, which may be some type of specialized smooth muscle cells peculiar to certain tissue, that are found in characteristic sites of the tumor.

Clinical Features

- *Age and sex*—it can occur at any age and females are affected more commonly than males.
- *Site*—most common site of occurrence is dorsum of tongue followed by skin, lips, breast, subcutaneous tissue, vocal cord and floor of mouth.
- *Appearance*—lesion which is found on tongue is usually single firm, submucosal nodule within the substance of the tongue itself.
- *Size*—the size of the tumor varies from a few millimeters to few centimeters.
- *Color*—color is usually pink but some tumor may appear yellow in color.
- *Signs*—lesion is not ulcerated and may have normal covering or may exhibit some clinical leukoplakia.

Diagnosis

- *Clinical diagnosis*—submucosal swelling on the dorsum of tongue may give clue to diagnosis (**Fig. 15-50**).
- *Laboratory diagnosis*—the lesion is composed of two types of cells: granular cells and satellite cells. The large granular cells are called as *Abrikossoff myocytes*.



Fig. 15-50: Submucosal nodule seen on tongue in granular cell myoblastoma.

Management

- *Surgical excision*—it is best treated by conservative local excision of the tumor.

Giant Cell Neoplasm

Central Giant Cell Tumor

It is the term applied for the lesions, which contain giant cells. It is chiefly a tumor of long bones, occurring at the epiphyseal end involving the adjacent metaphysis. It is an extremely uncommon tumor of head and neck and creates a lot of confusion with respect to diagnosis. Today these tumors represent benign tumors of osteoclastic origin.

Origin of Giant Cells

Different theories are given to describe the origin of giant cells in these lesions:

- *Resorption of deciduous roots*—Giant cells might be derived from the proliferating giant cells associated with the resorption of deciduous tooth roots. But for this, association of the lesion in transition period i.e. from of deciduous to permanent tooth should be there: but such association is found only in few cases.
- *Endothelial cells of capillaries*—another theory states that it originates from the endothelial cells of capillaries. To support this theory: there is a common occurrence of giant cells in vascular channels, suggesting that they arise from endothelial cells.
- *Modified form of cells*—Sapp found that giant cells, ultrastructurally, contain a sufficient number of features in common with osteoclasts. This concludes that they represent a slightly modified form of the cells.

Clinical Features

- *Age and sex distribution*—it is most frequently seen in 3rd and 4th decades, it is unusual in patients less than 20 years. Women are more affected as compared to males.
- *Site*—the commonest sites for giant cell tumor are the lower end of the femur, upper end of the tibia and lower end of the radius. In the oral cavity it is rare and if found it is in jaw.
- *Symptoms*—the principal symptoms are swelling of the bone accompanied in some cases by pain. The color of lesion is blue due to its cortical and mucosal thinning and internal vascularity (**Fig. 15-51**).
- *Signs*—the swelling may be tender and egg shell crackling can be elicited in large tumors. There is also expansion of jaw which is painless.



Fig. 15-51: Bluish color of central giant cell tumor shows its vascular nature (Courtesy Dr Tapasya Karamore).

Radiological Features

- *Radiodensity*—there is an area of radiolucency with thinning and expansion of the cortex, with little or no periosteal new bone formation.
- *Appearance*—the radiolucent area may show soap bubble or honeycomb appearance but this is inconsistent. Thinning of cortices is also present (**Fig. 15-52**).
- *Effect on teeth*—there may be resorption of roots of tooth. Displacement of teeth can also occur.

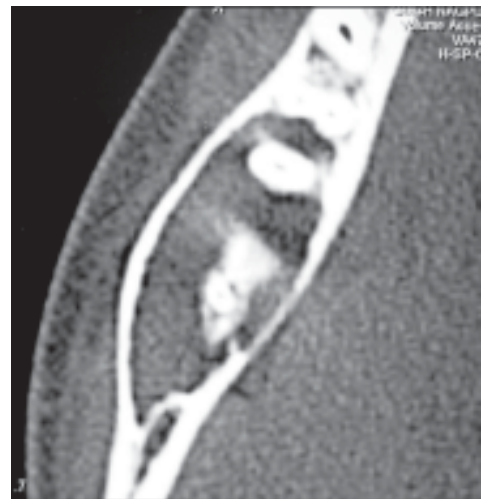


Fig. 15-52: Thinning of cortex seen in central giant cell tumor (Courtesy Dr Parate).

Diagnosis

- *Clinical diagnosis*—bluish color vascular lesion may suspect central giant cell tumor.
- *Radiological diagnosis*—multilocular lesion with thin cortex will give clue to diagnosis.
- *Laboratory diagnosis*—the tumor forms a maroon or reddish brown fleshy mass that replaces the spongiosa of the bone. It consists of numerous giant cells lying in cellular matrix composed of spindle shaped cells and scanty collagen.

Management

- *Curettage*—the usual method of treatment is curettage, but this is followed by a high recurrence rate.
- *Intralesional corticosteroid*—nowadays, intralesional injection of corticosteroids proves to be beneficial in many cases. Suggested use of triamcinolone 10 mg/ml of once in week for 6 weeks. 1 ml injection is given for each 1 cm of jaw involvement. Local anesthesia is added in the injection.

Peripheral Giant Cell Granuloma

It is also called as '*peripheral giant cell reparative granuloma*', '*giant cell epulis*', '*osteoclastoma*' and '*peripheral giant cell tumor*'. It is five times more common as compared to central giant cell granuloma. It seems to originate from either periodontal ligament or mucoperiosteum.

Etiology

- *Injury*—it is an unusual response of tissue to injury. The trauma may be caused by tooth extraction and dental irritation.
- *Chronic infection*—it can occur in chronic infections.
- *Hormonal*—it may appear under the stimulus of increased circulating parathormone i.e. primary and secondary hyperparathyroidism.

Clinical Features

- *Age and sex distribution*—most oftenly seen over 20 years of age, with an average of 45 years. Females are affected twice as common as males. Predominant in white persons.
- *Common site*—it occurs on gingiva and alveolar mucosa, most frequently anterior to molars. It is common in mandible than maxilla.
- *Early lesion*—it appears as discoloration and slight swelling of the buccal aspect of the gingiva.
- *Later lesion*—the lesion increases in size and becomes rounded and very often pedunculated (**Fig. 15-53**).
- *Hourglass appearance*—sometimes, it grows in an hourglass manner, with the waist of the lesion between two teeth and the globular extremities presenting buccally and lingually (**Fig. 15-54**).
- *Color*—the color of the lesion is usually dark red (**Fig. 15-55**) or maroon color. If sufficient amount of hemosiderin exists near the periphery, the lesion is bluish, otherwise it is red and/or pink. Lesions with much fibrous tissue are paler.
- *Consistency*—it may feel soft to hard.
- *Size*—the lesions vary in size from 0.5 cm to 1.5 cm in diameter.



Fig. 15-53: Rounded lesion of peripheral giant cell granuloma seen on gingiva.



Fig. 15-54: Peripheral giant cell granuloma showing hourglass appearance.



Fig. 15-55: Dark red color of peripheral giant cell granuloma seen in buccal sulcus.

- *Signs*—it is vascular or hemorrhagic and sometimes ulceration is also present. There may be tenderness on palpation.
- *Edentulous area*—in edentulous patients, it may present as a vascular, ovoid or fusiform swelling of the crest of the ridge, seldom over 1-2 cm in diameter or there may be granular mass of tissue which seems to be growing from the tissue covering the slope of the ridge.

Radiological Features

- *Edentulous area*—in edentulous areas, the peripheral giant cell granuloma characteristically exhibits superficial erosion of the bone with peripheral 'cupping' of the bone.
- *Dentulous area*—in dentulous areas, there may be superficial destruction of the alveolar margins or crest of the interdental bone.

Diagnosis

- *Clinical diagnosis*—dark red color lesion with hourglass appearance soft in consistency will favor the diagnosis of peripheral giant cell granuloma.
- *Laboratory diagnosis*—multinucleated giant cells and young connective tissue spindle shaped cells are scattered throughout the granulation tissue in delicate reticular and fibrillar connective tissue stroma.

Differential Diagnosis

- *Hemangioma*—present at birth, seldom occurs on gingiva.
- *Lymphangioma*—it is rare on gingiva and color is same as the tissue.
- *Metastatic carcinoma*—history of primary tumor.
- *Oral nevi and nodular melanoma*—firm on palpation and darker in color.

Management

- *Surgical excision*—excision with borders of normal tissue with entire base of the lesion, so that recurrence is avoided.
- *Removal of cause*—elimination of chronic irritant.

Teratoma

It is also called as 'teratoblastoma' or 'teratoid tumor'. It is a neoplasm of different types of tissue which are not native to the area in which the tumor occurs. True teratoma is developmental tumor which is composed of all the three germ layers ectoderm, mesoderm and endoderm. Out of these three layers, one of the layers exhibits neoplastic proliferation.

It is congenitally acquired and usually found in the ovary. The finding of various organs like structure i.e. teeth,

tissue, hair and skin are seen in this tumor. The lesion that arises in the base of the skull often extends into the cranial and oral cavity; newborn infants with such lesions rarely survive.

Clinical Features

- *Age*—it is known to be present at birth or is discovered shortly thereafter.
- *Site*—it can occur in ovaries, testes, anterior mediastinum, retroperitoneal area, presacral and coccygeal regions, pineal region, head, neck and abdominal viscera. In oral cavity it can be seen on hard palate and soft palate area.
- *Symptoms*—they are benign lesions and grow slowly.
- *Signs*—they often contain recognizable hair, sebaceous material and teeth.



A



B

Figs 15-56A and B: (A) Child patient with appearance of double palate. (B) Same patient after 2 years showing double arch and double palate (Courtesy Dr Debta).

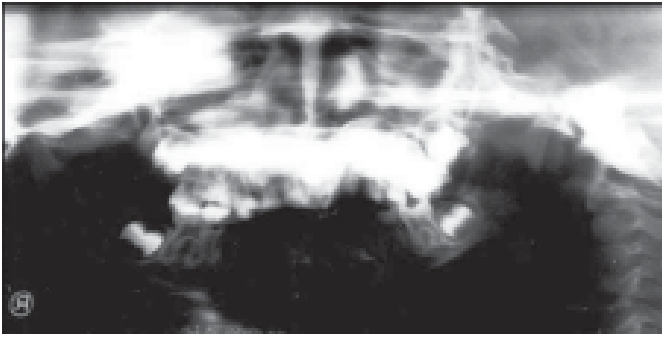


Fig. 15-57: Radiograph of same patient showing double arch and double palate appearance (Courtesy Dr Debta).

- *Epignathus*—true teratoma occurs sometimes and it is called as '*epignathus*'. It arises from the hard and soft palate which may be perforated. The tumor tends to fill the buccal cavity and may extend into the mouth. Epignathi may develop in the region of Rathke's pouch and may be found entirely above the palate or even within the cranium. In rare cases, grapefruit size of tumor protruding from the mouth may arise from the palate and can be a well formed finger with a finger nail on its upper part.
- *Double palate*—this is a rare type of teratoma which occurs in hard palate area. There is occurrence of double palate with double arch in the palate area (**Figs 15-56 A and B**). Double palate can also be seen on panoramic radiograph (**Fig. 15-57**).

Diagnosis

- *Clinical diagnosis*—it is not difficult to make clinical diagnosis as tissue present in teratoma is not of that particular site.
- *Laboratory diagnosis*—biopsy shows epithelium and epithelial appendages such as hair, sweat gland, sebaceous glands, salivary glands and teeth as well as discrete epithelial organs such as thyroid gland and pancreas.

Management

- *Surgical*—surgical removal of the tumor should be carried out.

Salivary Gland Tumor

It is discussed in Chapter 26: *Salivary Gland Disorders*.

Inflammatory Hyperplasia

Chronic injury produces inflammation, which in turn stimulates the formation of granulation tissue that consists of proliferative endothelial tissue with rich patent

capillary bed, chronic inflammatory cells and few fibroblasts. They are smoothly contoured or lobulated with very red appearance due to rich vascularization and transparency of non-keratinized epithelial covering. It has got soft, spongy and broad sessile base.

Pyogenic Granuloma

It is also called as '*granuloma pyogenicum*'. It is a response of tissues to non-specific infection of staphylococcus or streptococci. It is relatively a common soft tissue tumor of the skin and mucous membrane. It is known to be a reactive inflammatory process in which there is an exuberant fibrovascular proliferation of the connective tissue, secondary to some low grade chronic irritant.

Etiology

- *Microorganisms*—it is usually due to botryomycotic infection, staphylococci and streptococci.
- *Trauma*—it may occur as a result of minor trauma to the tissue, which provides a pathway for the invasion of tissues by non-specific of microorganisms.
- *Local irritant*—it is an inflammatory response to local irritation such as calculus.
- *Hormonal imbalance*—it is a contributing factor for the development of some pyogenic granulomas.

Clinical Features

- *Age and sex*—females are affected more than males with common age of occurrence are 11 to 40 years.
- *Sites*—common sites are gingiva, lip, tongue, buccal mucosa, palate, vestibule and alveolar mucosa.
- *Symptoms*—it is an asymptomatic papular, nodular polypoid mass.
- *Appearance*—lesions are elevated, pedunculated or sessile masses with smooth, lobulated or even warty surface.
- *Surface*—surface can commonly ulcerate and shows tendency to hemorrhage upon slightest pressure or trauma.
- *Pus*—sometimes, there is exudation of purulent material.
- *Color*—it is deep red to reddish purple depending on the vascularity (**Fig. 15-58**); it is painless and rather soft in consistency, with some lesions having brown cast, if hemorrhage has occurred into the tissues. If lesion is of mixed variety it appears red with pink areas and if it is completely fibrous it is slightly pink and firm on palpation.
- *Consistency*—it feels moderately soft and bleeds readily.
- *Size*—it may develop rapidly, reaching full size and remain static for an indefinite period with size ranging from few millimeters to centimeters. Average size of lesion being 0.9 to 1.2 cm.



Fig. 15-58: Pyogenic granuloma presenting as reddish purple mass on gingiva (Courtesy Dr Chole).

Diagnosis

- *Clinical diagnosis*—reddish purple lesion, soft in consistency can be pyogenic granuloma.
- *Laboratory diagnosis*—biopsy shows granulation tissue which is exuberant and is usually localized. Overlying epithelium, if present, is generally thin and atrophic, but may be hyperplastic.

Differential Diagnosis

- *Small benign and malignant mesenchymal tumor*—higher incidence of pyogenic granuloma.
- *Exophytic capillary hemangioma with ulceration*—most of the hemangiomas are blanched on pressure while pyogenic granuloma does not blanch on pressure.
- *Peripheral giant cell granuloma*—lesion is more bluish, as compared to pyogenic granuloma which is red to pink. There is also radiological evidence of superficial cuffing of the alveolar bone.
- *Peripheral fibroma with calcifications*—underexposed radiograph shows small radiopaque foci within the shadow of growth.

Management

- *Surgical excision*—surgical excision is done and care is taken to remove the calculus of adjacent teeth as it may act as a local irritant and cause recurrence.
- *Removal of irritant*—elimination of the causative agent should be done.

Pregnancy Tumor

Pregnancy tumor usually develops in first trimester of pregnancy. The size usually increases in 7 months of pregnancy (Fig. 15-59).



Fig. 15-59: Pregnancy tumor present as growth in upper maxillary lesion (Courtesy Dr Abhishek Soni).

Etiology

- *Local minor trauma*—it occurs as a result of local minor trauma and in cases where tissue reaction is intensified by endocrine alteration occurring during pregnancy.

Clinical Features

- *Age*—it occurs during pregnancy, often in about 3rd month of pregnancy and sometimes later.
- *Appearance*—it is a well defined lesion, which gradually increases in size (Fig. 15-60).
- *Regression after pregnancy*—maximum cases of pregnancy tumor regress after pregnancy. If surgically removed during pregnancy, it usually recurs.



Fig. 15-60: Well defined lesion seen in 3rd trimester of pregnancy (Courtesy Dr Abhishek Soni).

Radiological Appearance

- *Rarefaction of bone*—in some of the cases there is slight rarefaction of the bone beneath the attachment of the tumor, usually at the crest of the alveolus.

Diagnosis

- *Clinical diagnosis*—well defined tumor-like lesion in pregnancy will diagnose this condition.
- *Radiological diagnosis*—not so specific.
- *Laboratory diagnosis*—biopsy shows similar feature as that of pyogenic granuloma.

Management

- *No treatment*—no treatment should be given to patient as maximum cases regress by its own after pregnancy.
- *Surgical excision*—if pregnancy tumor does not resolve after pregnancy, it should be surgically excised.

Parulis

It is a small inflammatory hyperplastic type of lesion that develops on the alveolar mucosa at the oral terminal of draining sinus (**Fig. 15-61**). Maxillary labial and buccal mucosa are commonly affected. Slight digital pressure on periphery of parulis may force a drop of pus from the sinus opening. It usually regresses after the infection is eliminated.



Fig. 15-61: Parulis presented as hyperplastic growth at the sinus opening of maxillary premolar region.

Inflammatory Papillary Hyperplasia

It is also called as 'palatal papillomatosis' and 'palatal epithelial hyperplasia'. It occurs in 3% to 4% of dentures wearers.

Etiology

- *Frictional irritation*—it is produced by loose fitting dentures on palatal tissues. It occurs in patients who sleep with their dentures. It is much common in acrylic dentures than in those with metallic dentures.

- *Suction chamber in palatal seating*—full dentures in which relief areas or suction chambers are cut in palatal seating surface appear to be the strongest stimuli for the lesion.
- *Poor denture hygiene*—it is more common in patients with poor oral hygiene.
- *Amalgam filling irritation*—in some cases, irritation due to amalgam filling may produce these lesions.

Clinical Features

- *Age and sex*—It can arise at any age in adults and has no definite sex predilection.
- *Site*—it occurs exclusively on palate beneath the complete or partial denture. It occurs predominately in edentulous patients and the site of lesion corresponds to the denture base. In rare cases, cheek may be involved.
- *Appearance*—whole palatal mucosa under the denture may be covered with numerous small polypoid masses.
- *Warty appearance*—the lesion presents as numerous closely arranged, red, edematous papillary projections (**Fig. 15-62**) often involving nearly all of the hard palate and imparting it to a 'warty appearance'.



Fig. 15-62: Inflammatory papillary hyperplasia showing papillary projection (Courtesy Dr Bande).

- *Overripe berry appearance*—in some cases, it is swollen and papillary projection resembles the surface of 'overripe berry'.
- *Cobblestone appearance*—in some cases, it produces a 'cobblestone' appearance (**Fig. 15-63**).
- *Signs*—lesions are friable, often bleed with minimum trauma and may be covered with thin whitish exudate. The tissue exhibits varying degrees of inflammation, but seldom there is ulceration.
- *Size*—these are seldom over 0.3 cm in diameter. The lesion may extend onto the alveolar mucosa. The individual papillae are seldom over a millimeter or two in diameter.

- *Candida infection*—when complicated by *Candida albicans*, lesion appears as red to scarlet, soft and bleeds easily in the inflammatory or granulomatous stage.



Fig. 15-63: Palatal papillary hyperplasia showing cobblestone appearance.

Diagnosis

- *Clinical diagnosis*—papillary projection beneath the palatal denture will diagnose this lesion.
- *Laboratory diagnosis*—biopsy shows numerous small vertical projections, each composed of parakeratotic or sometimes orthokeratotic stratified squamous epithelium and a central core of connective tissue.

Differential Diagnosis

- *Nicotina stomatitis*—Occurs almost exclusively in pipe smokers who are not wearing maxillary complete dentures. Lesions are more nodular and broader but less elevated. Red dot is seen in center.
- *Verrucous carcinoma*—papillary hyperplasia may mimic verrucous carcinoma. But here the papillary hyperplasia is not associated with denture. Biopsy should be done to rule out verrucous carcinoma.

Management

- *Removal of denture*—remove the denture at night to provide rest to the tissue. In many cases tissue will resume normal appearance after denture is removed.
- *Conditioning liner*—conditioning liner should be applied under the denture. This will act as palatal dressing and promotes greater comfort.
- *Topical antifungal treatment*—in case with superimposed candidal infection, topical application of antifungal agents should be used. Most commonly used is nystatin ointment.

- *Surgical removal*—it is done by partial thickness or full thickness surgical blade excision, curettage, electrosurgery and cryosurgery. This is done in case of fibrosis of the tissue.

Epulis Granulatosum

It is reactive hyperplasia that develops within a tooth socket after the extraction or exfoliation of tooth. It is caused by sharp spicules of bone in socket.

Clinical Features

- *Age*—it can occur at any age and becomes apparent within 2 weeks after the loss of a tooth.
- *Appearance*—it is exuberant dark red granulation tissue extruding from the socket (Figs 15-64A and B).
- *Symptoms*—it is painless growth.
- *Signs*—the enlargement is soft, hemorrhagic with an erythematous to white, smooth surface.



Figs 15-64A and B: Epulis granulatosum showing growth in the extraction socket.

Diagnosis

- *Clinical diagnosis*—hyperplasia in extraction socket will be diagnosed this condition.

Differential Diagnosis

- *Antral polyp*—sinus wall defect is usually demonstrated either radiographically or by requesting the patient to blow air through the nose while nostrils are occluded.
- *Pulp polyp*—extensive destruction of tooth can be seen.
- *Malignancy*—radiological changes are present.

Management

- *Curettage*—it is done to remove the granulation tissue and smoothing of the socket borders is indicated.
- *Excisional biopsy*—it should be done.

Nodular Fasciitis

It is also called as '*pseudosarcomatous fasciitis*'. It is non-neoplastic connective tissue proliferation. It histologically mimics malignant mesenchymal neoplasm but its clinical behavior is benign. It appears to be an inflammatory reactive phenomenon.

Clinical Features

- *Age and sex distribution*—it is more commonly seen in 4th and 5th decade of life. There is no sex predilection.
- *Site*—it is more common on the trunk and extremities. Intraorally, it is seen on buccal mucosa followed by subcutaneous tissues overlying the mandible, zygoma, parotid sheath and oral mucosa.
- *Symptoms*—it appears as a small lump, which may be painful.
- *Sign*—it has got exophytic presentation.
- *Size*—the lesion often enlarges rapidly, but only to maximum size of 4 cm, where it remains stationary or regresses.

Diagnosis

- *Clinical diagnosis*—it is not specific.
- *Laboratory diagnosis*—biopsy shows proliferation of fibroblast in haphazard manner in a vascular myxoid matrix which is rich in acid mucopolysaccharide. It results in a loose textured feathery appearance. There is frequent patchy chronic inflammatory infiltration, consisting of lymphocytes and occasional plasma cells and macrophages.

Management

- *Surgical excision*—local excision will yield excellent result.

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16

Malignant Tumor of Jaw

Oral Carcinoma

Oral carcinoma is one of the most prevalent cancers and is one of the 10 major causes of death. Most common oral carcinoma is squamous cell carcinoma. It is a disease of increasing age with 95% cases in people older than 40 years of age.

Cancer is a commonly used term for all malignant tumors. In Latin language, cancer means 'crab' which has a fat meaty body and mass extension which springs and invades the surrounding tissues.

Cancer may be defined as an uncontrolled tissue growth in susceptible patient, which results from imbalance between cell division and programmed cell death.

Tumor is an autonomous new growth of tissue or it is an abnormal mass of tissue, the growth of which exceeds and is uncoordinated with that of normal tissue and persists in

the same excessive manner even after the cessation of stimuli which evoked the changes.

It is classified in **Table 16-1**.

Characteristics of Malignant Lesion

- *Lesion borders*—they exhibit ill defined borders which are very irregular and ragged.
- *Rapid growth*—it increases in size very rapidly.
- *Metastasis*—they always metastasize either by direct spread, by lymphatics or through the bloodstream.
- *Adjacent cortical bone*—grows by invasion and causes destruction of adjacent structures. Bony cortex will be destroyed rather than expanded.
- *Radiodensity*—radiolucent or may be mixed with radiopacity.
- *Dental involvement*—root intact and tooth in position.

Table 16-1: Classification of malignant tumor

Epithelial • Squamous cell carcinoma • Metastatic carcinoma • Basal cell carcinoma • Transitional cell carcinoma • Malignant melanoma • Verrucous carcinoma • Spindle cell carcinoma • Primary intra-alveolar carcinoma • Intraepidermoid carcinoma • Adenoid squamous cell carcinoma Fibrous connective tissue • Fibrosarcoma • Malignant fibrous histiocytoma Adipose tissue • Liposarcoma Cartilage • Chondrosarcoma Bone • Osteosarcoma • Osteochondrosarcoma • Ewing's sarcoma Vascular • Hemangioendothelioma • Angiosarcoma	Neural tissue • Neurosarcoma • Neurofibrosarcoma • Neuroblastoma • Ganglioneuroma Muscle • Leiomyosarcoma • Rhabdomyosarcoma • Malignant granular cell myoblastoma Lymphoid tissue • Hodgkin's lymphoma • Non-Hodgkin's lymphoma • Primary reticular cell sarcoma • Burkitt's lymphoma • Leukemia Myeloma • Multiple myeloma • Plasmacytoma Tumor of salivary gland • Mucoepidermoid carcinoma • Adenocystic carcinoma • Adenocarcinoma • Acinic cell carcinoma • Malignant changes in pleomorphic adenoma
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Etiology and Risk Factors for Oral Cancer (Fig. 16-1)

Tobacco Use

Smokeless tobacco

- *Major etiological factors*—tobacco use is the major etiological factor for the development of oral cancer. The effect of tobacco use, heavy alcohol consumption and poor diet can probably explain over 90% of cases of oral cancer. Much of tobacco in the world is consumed without combustion, by being placed in contact with mucous membrane, through which nicotine is absorbed.
- *Why it causes oral cancer*—it contains potent carcinogens like nitrosamine, polycyclic hydrocarbons and polonium and metabolites of these constituents, which have been suggested etiologic factors in oral cancer.
- *Common forms of oral smokeless tobacco*—it includes *pan/ paan/betal quid* (it contains areca nut, betel leaf, slaked lime, catechu, condiments with or without tobacco), *khaini* (contains tobacco and lime), *mishri* (burned tobacco), *zarda* (boiled tobacco), *gadakhu* (tobacco and molasses), *mawa* (tobacco, lime and areca), *nass* (tobacco, ash, cotton or sesame oil), *naswar/niswar* (tobacco, lime, indigo, cardamom, oil and menthol), *shammah* (tobacco, ash and lime) and *toombak* (tobacco and sodium bicarbonate).

Smoking

- *Why it causes oral cancer*—tobacco smoke contains carbon monoxide. It is an important factor in the development of oral cancer. Study shows that cigar and pipe smoking increase the risk of cancer than cigarette smoking.
- It has been stated that the pooling of carcinogens in saliva gives rise to cancer in the floor of mouth and ventral and lateral tongue.
- Smoking is strongly associated with soft palate cancer than anterior sites.

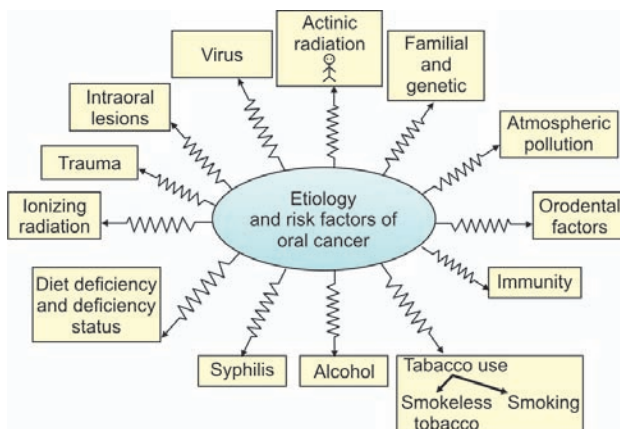


Fig. 16-1: Etiology of oral cancer diagrammatic representation.

Alcohol

- *Forms of alcohol*—all forms of alcohol; including hard liquor, wine and beer has been implicated in the etiology of oral cancer.
- *Mechanism*—the mechanism by which alcohol affects includes the dehydrating effect of alcohol on the mucosa which increases mucosal permeability and the effects of carcinogens on the mucosa. Beverage congeners include nitrosamines and impurities which can act as carcinogens.
- *Associated habit*—most of the heavy alcohol consumers use tobacco, so it is very difficult to separate the ill effects individually.

Actinic Radiation

It is a minor etiologic factor in case of lip cancer. Lip cancer occurs more commonly in fair skinned people who are generally engaged in outdoor occupation, such as farming and fishing.

Familial and Genetic

There is little evidence that there is familial and genetic predisposition for the development of oral cancer.

Lip cancer is amongst the sites which show strongest cancer clustering within families. But it also reflects the fact that families tend to have same occupation, i.e. fishing and farming, which is related with ultraviolet exposure.

Oral cancers are more prevalent in blacks as compared to white.

Atmospheric Pollution

Parts of the urban/rural difference in incidence of head and neck cancers have been related to atmospheric pollution.

Sulphur dioxide and smoke concentration in the atmosphere are positively correlated with squamous cancer of larynx and pharynx. The impact on cancer of the mouth is likely to be less, but merits careful study.

Blue collar workers exposed to dust or inhalation of organic and inorganic agents are at increased risk of cancers of mouth.

Orodental Factors

It is more prevalent in patients with poor oral hygiene, faulty restorations, sharp teeth, ill fitting dentures and those with syphilitic glossitis.

Vitamin A Deficiency

Vitamin A deficiency can produce excessive keratinization of the skin and mucous membrane. So their deficiency can predispose to the formation of carcinoma.

Immunity

The increasing incidence of oral cancers is clearly age related, which may reflect declining immune surveillance with age.

It may occur in immunosuppressed patients following organ and bone marrow transplantation. HIV/AIDS patients are at increased risk of oral cancer.

Syphilis

It is traditionally associated with oral cancer. Oral malignancy is commonly found in tertiary stage of syphilis. Another aspect why syphilis can lead to malignancy is that in the management of syphilis previously we use arsenic and heavy metal which has potential carcinogen.

Diet Deficiency and Deficiency Status

Nutritional deficiency and liver dysfunction can also play a role in it. The relationship between sideropenic dysphagia and oral cancer is well established.

Ionizing Radiation

Carcinoma of buccal mucosa may occur as a complication of long-term radiotherapy.

Trauma

Trauma in combination with other factors like chronic cheek biting, denture use and irregular teeth may act as a co-carcinogen and may promote transformation of epithelial cells.

Virus

The possibility of type I herpes simplex virus being associated with oral cancer has been suggested.

Other viruses which can lead to oral cancer are human papillomavirus and human immunodeficiency virus.

Intraoral Lesions

- *Chronic ulceration and fissure*—it can cause oral cancer particularly of lip.
- *Lichen planus*—reports of carcinoma supervening on lichen planus have been made in some cases.
- *Candidiasis*—it is possible precursor of carcinoma. Candidal infection is often associated with acanthosis and parakeratosis.
- *Leukoplakia*—it is common pre-malignant condition seen in oral cavity. Nodular leukoplakia show higher rates of epithelial dysplasia.
- *Median rhomboid glossitis*—in some cases, it has followed by cancer.

- *Plummer Vinson's syndrome*—the atrophic changes in the mucous membrane of the upper alimentary tract that occur in this syndrome may predispose to malignancy.
- *Submucous fibrosis*—it is precancerous condition occurs in oral cavity.
- *Oral melanosis*—it appears to be associated with oral cancers in India.
- *Discoid lupus erythematosus*—a number of cases of carcinoma of lip developing in the lesion of lupus erythematosus have been reported.
- *Epidermolysis bullosa*—it is occasionally followed by carcinoma.

Epithelial Tumors

Peripheral Squamous Cell Carcinoma

It is also called as '*epidermoid carcinoma*' or '*epithelioma*'. It represents 90% of all malignant tumors occurring in the mouth and jaws. The majority of oral cancer cases are of squamous cell carcinoma. The oral lesion often invades the jaw. It arises in gingival tissue, buccal sulcus, floor of mouth and some other portion of oral mucosa.

Clinical Features

General features

- *Age and sex*—predominately, it occurs in males with ratio of 2:1, older than 50 years with an average age of approximately 60 years.
- *Site*—most commonly involved are the posterior and lateral borders of the tongue and lower lip and less frequently the floor of mouth, alveolar mucosa, palate and buccal mucosa. It may be solitary and multifocal.
- *Symptoms*—patient may present with awareness of a mass in the mouth and neck. Small lesion is asymptomatic. Large lesions may cause some pain or paresthesia and swelling. Patients complain of persistent ulcer in the oral cavity. Function of organ in which malignancy occur is impaired.
- *Appearance*—the clinical appearance of a carcinomatous ulcer is that one of irregular shape (**Fig. 16-2**), indurated and raised everted edges.
- *Base*—usually have broad base and are dome-like or nodular. Base is firm on palpation.
- *Exophytic lesion*—it has irregular, fungating, papillary and verruciform surface. The surface is ulcerated and base is hard on palpation (**Fig. 16-3**).
- *Endophytic lesion*—this is depressed irregularly shaped ulcerated central area with surrounding rolled border. The rolled border results from invasion of tumor in the tissue (**Fig. 16-4**).



Fig. 16-2: Irregular shaped ulceration seen in buccal vestibule due to malignancy (Courtesy Dr Chole).



Fig. 16-4: Endophytic type of growth seen in carcinoma of palate which shows rolled border (Courtesy Dr Suwas Darvekar).



Fig. 16-3: Exophytic growth seen in malignancy showing irregular margins and surface ulceration (Courtesy Dr Suwas Darvekar).



Fig. 16-5: Surface of oral malignancy is whitish gray and pebbly (Courtesy Dr Parate).

- **Surface**—surface may range from granular to pebbly to deeply creviced (**Fig. 16-5**). In some cases, surface may be entirely necrotic and have ragged whitish gray appearance.
- **Color**—it may be completely red or red surface may be sprinkled with white necrotic or keratin area.
- **Lymph nodes**—superficial and deep cervical nodes are commonly affected. They become enlarged and are firm to hard on palpation. The nodes are non-tender unless associated with secondary infection or an inflammatory response. It may be nodular or polypoid and pink to red and have at least one ulcerated patch on their surface. Fixation of nodes to adjacent tissues occur later.

- **Effect on adjacent tissues**—fixation of primary tumor to adjacent tissues, i.e. overlying bone suggests involvement of periosteum and possible spread to bone.
- **Field cancerization**—tendency of development of multiple mucosal cancers, is called as field cancerization. This occur due to diffuse exposure of local carcinogen.

Carcinoma of floor of mouth

- **Site**—most frequently in the anterior portion of floor.
- **Appearance**—the typical carcinoma of the floor of mouth is an indurated ulcer of varying size, on one side of the midline (**Fig. 16-6**). It may take form of wart-like growth, which tend to spread superficially rather than in depth.
- **Symptoms**—it may or may not be painful. In some cases, there may be referred pain in the ears. The proximity of this tumor to the tongue produces some restricted/limited movements of tongue, often induces peculiar thickening



Fig. 16-6: Carcinoma involving floor of mouth showing indurated ulceration.

or slurring of the speech. There may be excessive salivation.

- *Teeth mobility*—carcinoma in close relation to teeth may cause loosening or exfoliation and root resorption.
- *Extent*—carcinoma of floor of mouth may invade the deeper tissues and may even extend into the sub-maxillary and sublingual glands.
- *Metastasis*—metastasis from the floor of the mouth are found most commonly in the sub-maxillary group of lymph nodes and since the primary lesion frequently occurs near the midline where lymphatic cross drainage exists, contralateral metastasis is often present.

Carcinoma of buccal mucosa (Figs 16-7A and B)

- *Site*—the lesions develop most frequently along or inferior to a line opposite the plane of occlusion. It usually occurs opposite to the third molar.
- *Symptoms*—the lesion is often painful.
- *Appearance*—the tumor begins as small nodules and enlarges to form a wart-like growth which ultimately ulcerates (**Fig. 16-8**).
- *Extent*—there is induration and infiltration of deeper tissues. Extension into the muscle of neck, alveolar mucosa and ultimately into bone may occur.
- *Exophytic growth*—some cases appear to be growing outward from the surface rather than invading the tissues is called as exophytic or verrucous growth.
- *Metastasis*—the most common site of metastasis is the submaxillary lymph nodes.

Carcinoma of labial mucosa

- *Cause*—it is frequently encountered in person who habitually keeps a mixture of tobacco lime in the labial vestibule.
- *Site*—the lower labial mucosa is more commonly involved than the upper.
- *Symptoms*—the most common initial signs and symptoms are growth or swelling, soreness and ulceration (**Fig. 16-9**).



Figs 16-7A and B: Carcinoma of buccal vestibule which also involve buccal mucosa (Courtesy Dr Suwas Darvekar).



Fig. 16-8: Carcinoma of buccal mucosa showing ulceration and white surface in the lesion (Courtesy Dr Parate).



Fig. 16-9: Carcinoma labial sulcus showing extensive lesion and ulceration.

- *Extent*—advanced lesion may be ulcerative-infiltrative type, showing exophytic growth.
- *Lymph nodes*—lymph node involvement may occur, which may be unilateral or bilateral (**Fig. 16-10**).



Fig. 16-10: Carcinoma of labial sulcus showing exophytic growth (Courtesy Dr Suwas Darvekar).

Carcinoma of palate

- *Cause*—it is common in area where reverse smoking is practiced.
- *Sex*—it is seen more commonly in females as compared to men, in case of reverse smoking.
- *Appearance*—palatal cancer usually manifests as a poorly defined ulcerated painful lesion on one side of the midline (**Figs 16-11A and B**).
- *Base and surface*—most of the lesions are exophytic and with broad base and nodular surface.



Figs 16-11A and B: Carcinoma of palate showing extensive lesion and ulceration (Courtesy Dr Suwas Darvekar).

- *Extent*—it frequently crosses the midline and may extend laterally to include tonsillar pillars or even the uvula. The tumor of hard palate may invade the bone or occasionally the nasal cavity. While infiltrating, lesions of the soft palate may extend into the nasopharynx.

Carcinoma of oropharynx

- *Site*—carcinoma of soft palate and oropharyngeal mucosa can occur.
- *Symptoms*—dysphagia, i.e. difficulty in swallowing is the most common complaint. Patient may complain of pain which is dull and sharp and is referred to ear.
- *Appearance*—it is same as that of other carcinoma.
- *Size*—size of the tumor is always greater than other carcinoma of the oral cavity.

Other specific carcinomas of gingiva, tongue, lip and maxillary sinus are described in their respective chapters.

Radiographic Features

- *Site*—it may be found along the entire border of the tumor or may be restricted to a relative small area. Irregular spicule of bone may be left behind.

- *Appearance*—there is roughly semicircular or saucer shaped erosion into the bone surface (Fig. 16-12).

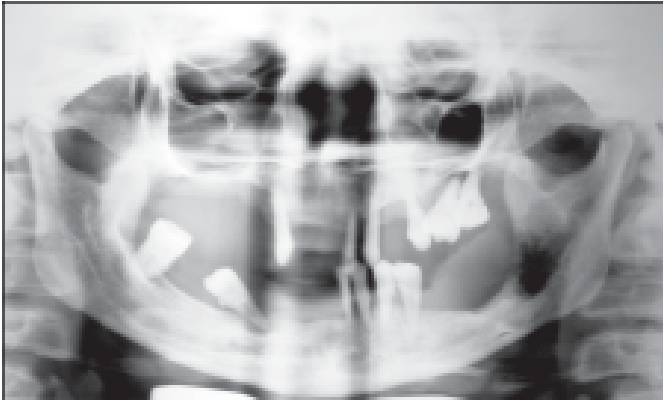


Fig. 16-12: Saucer shaped erosion of mandible in lower left posterior region.

- *Margins*—there are ragged ill-defined borders that illustrate the varying uneven osteolytic invasion (Fig. 16-13). Rarely, the border may appear smooth without cortex indicating underlining erosion rather than invasion.



Fig. 16-13: Ill-defined margin seen in case of malignancy of maxillary posterior region (Courtesy Dr Parate).

- *Finger-like extension*—if the bone involvement is extensive then periphery appears to have finger-like extensions preceding a zone of impressive osseous destruction (Fig. 16-14).
- *Infiltration of bone*—little 'bays' of bone destruction extend into the bone, leaving irregular promontories of variable length and width extending into the main area of bone destruction. Margins of 'bays' are irregular and jagged. These produce a finger-like projection and this appearance is described as infiltration of the bone.
- *Pathological fracture*—sometimes when it involves the lower border of mandible, there may be so much bone loss where pathological fracture occurs (Figs 16-15 and 16).



Fig. 16-14: Finger-like extension seen in mandible in squamous cell carcinoma.

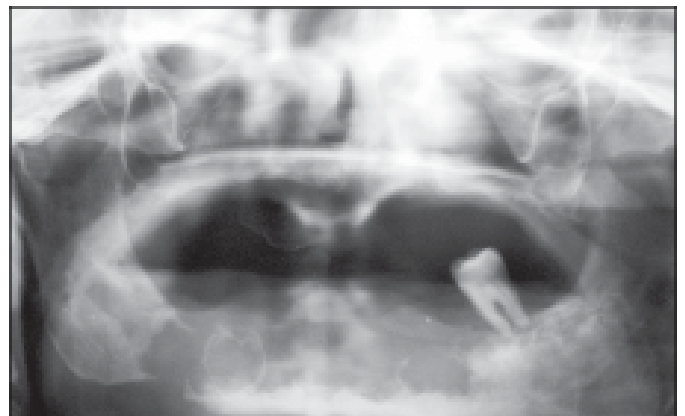


Fig. 16-15: Carcinoma involving mandible with pathological fracture on right side.

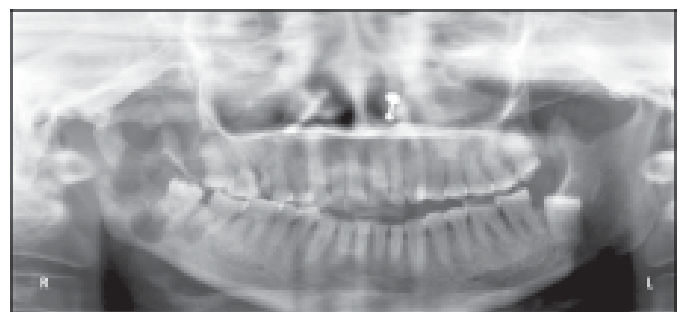


Fig. 16-16: Squamous cell carcinoma in parotid region with fracture.

- *Soft tissue mass*—a soft tissue mass is often present with carcinoma of jaw and this may be seen as faintly increased density in the radiograph (Fig. 16-17), standing above the general level of the bone.
- *Effect on surrounding tissues*—tumors may grow along the inferior neurovascular canal and through the mental foramen, resulting in an increase in the width and

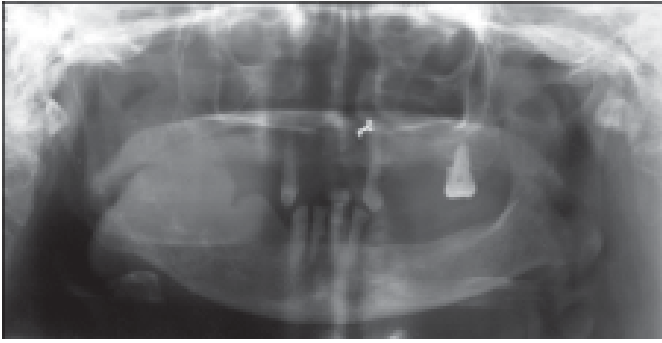


Fig. 16-17: OPG showing soft tissue shadow which is seen as faintly increased density (Courtesy Dr Parate).

eventually the loss of cortical boundaries. Destruction of floor of nose, maxillary sinus and buccal or lingual mandibular plate may occur. The inferior border of mandible may be thinned or destroyed.

- *Effect on teeth*—there is widening of the periodontal ligament space with loss of adjacent lamina dura. Teeth appear to float in a mass of radiolucent soft tissue deprived of any bony support (**Fig. 16-18**). In extensive tumor soft tissue mass may grow into the teeth and teeth appear to be grossly displaced from its normal position.

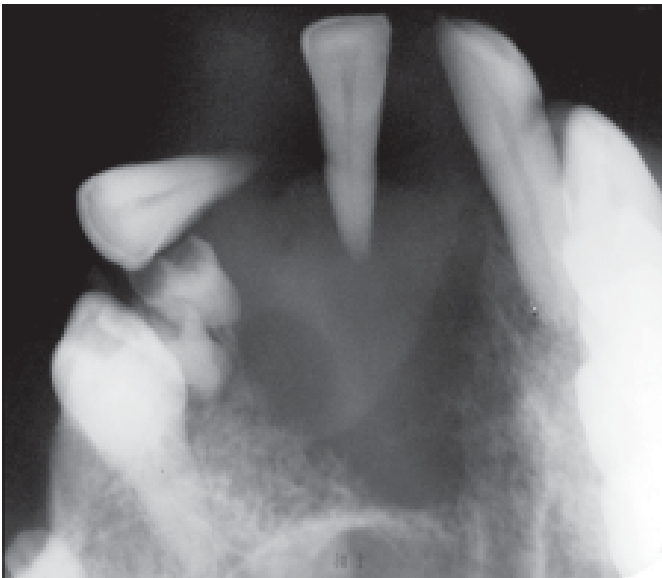


Fig. 16-18: 'Floating teeth appearance' seen in maxillary anterior region.

- *Carcinoma of floor of mouth and buccal sulcus*—they cause bone destruction from sides of bone. In some cases, there is radiolucent area which contains small islands of slightly greater density. These areas represent finger-like projection of unaffected bone situated between the bay-like areas of infiltration and bone destruction, with the rays passing through the length of finger-like processes.

Diagnosis

- *Clinical diagnosis*—ulcerative growth with indurated base, rolled margin goes in favors of squamous cell carcinoma.
- *Radiological diagnosis*—ill defined radiolucency with infiltrative margin may diagnose this condition.
- *Laboratory diagnosis*—in this, there is increased number of mitotic figures, hyperchromatism with prominent and multiple nucleoli and increased nucleocytoplasmic ratio. There is also pleomorphism of cells, keratinization, and keratin pearls deep to epithelial surface and loss of intercellular bridges. Histopathologically, it can be well differentiated, moderately differentiated and poorly differentiated.

Differential Diagnosis

Clinical

- *Ulcerated benign tumor*—ulcer should heal in one to two weeks after removal of mechanical noxious agent.
- *Basal cell carcinoma*—a history of slower development and more common due to exposure to sunlight.
- *Ulceration due to systemic diseases*—usually multiple, generalized poor condition and is usually painful.
- *Primary syphilitic lesion*—causative agent is known, no pain, indurated ulcer usually reddish brown with copper color halo.
- *Necrotizing sialometaplasia*—rare inflammatory lesion of smaller salivary glands, particularly in the posterior hard palate, ulcers are usually painful with no raised borders, no hardening and characteristic histology.
- *Peripheral metastatic tumor*—low frequency.
- *Malignant salivary gland tumor*—common on posterior hard palate, common in women and maintain their dome-shape appearance.
- *Verrucous carcinoma*—slow growing and is associated with prolonged use of tobacco, surface is papillomatous and white due to retention of keratin.
- *Pyogenic granuloma*—moderately soft to palpation and bleed easily.

Radiological

- *Osteomyelitis*—it usually produces some periosteal reaction whereas squamous cell carcinoma does not.
- *Osteoradionecrosis*—history of radiation therapy and prior malignancy.
- *Periodontitis*—the margins of periodontitis are smooth and well defined as compared to carcinoma. Periodontitis is usually generalized and if it is localized, there is specific cause for it.
- *Papillon Lèfevre syndrome*—it produces symptoms of infection and shows areas of sequestration.

Management

Management is discussed in detail at the end of this chapter:

- *Removal of cause*—removal of any local irritants i.e. the (5-S) smoking, spices, spirit, sepsis and syphilis.
- *Excisional biopsy*—excisional biopsy, if lesions are small.
- *Surgery*—cryosurgery, laser surgery and radical surgery.
- *Radiation*—radiation can be given in squamous cell carcinoma.
- *Chemotherapy*—chemotherapy is usually given in patient who cannot undergo surgical or radiation therapy.

Metastatic Carcinoma

It is also called as 'secondary carcinoma'. It is the most common malignant tumor in the skeleton. This tumor is transported to an area distant from its origin and establishes a new foothold and are said to have metastasized. Although the metastatic carcinoma of jaw is uncommon, its recognition is important because the jaw tumors may be the first indication that the patient has a malignant disease. Oral diagnostician can make an invaluable contribution of pathologic process of bone. Most common sites of origin are breast, lung, kidney, thyroid, prostate and colon.

Clinical Features

- *Age*—it is found in patients between 40 and 60 years of age and there may be history of primary tumor.
- *Site*—mandible is involved much more frequently than maxilla (**Fig. 16-19**), especially in the region near premolars and molars as tumor metastasizes to those bones which are rich in hemopoietic marrow. It is said that blood flow rate is decreased in areas of



Fig. 16-19: Extraoral swelling seen in metastatic carcinoma in mandibular region.

hemopoietic marrow and this predisposes tumor emboli to settle and grow in these areas. This hypothesis is consistent with jaw metastases as hemopoietic marrow is most routinely found at this site. The other sites involved are maxillary sinus, anterior hard palate and mandibular condyle.

- *Symptoms*—there may be pain, swelling, and paresthesia.
- *Numb chin syndrome*—it occurs when metastatic carcinoma involve inferior alveolar nerve. There is unexplained loss of sensation in lower lip and chin.
- *Appearance*—early lesion is nodule or dome shaped with smooth surface and due to trauma, may get ulcerated.
- *Teeth mobility*—teeth in this region may become loose or exfoliate and root resorption may occur. There is loss of bony support with periodontitis.
- *Swelling*—on occasion, tumor may breach the outer cortical palate of jaws and extend into surrounding soft tissue or presents as an intraoral mass.
- *Unhealed socket*—metastatic tumors are diagnosed only when the sockets of extracted teeth do not heal because of periodontal disease.
- *Loss of function of muscle*—if invasion occurs in muscle then their functions are impaired.
- *Pathological fracture*—there may be pathological fracture of the jaw or hemorrhage from the tumor site.

Radiographic Features

Osteolytic type

- *Radiodensity*—ill-defined radiolucent destructive lesion may be single or multiple and vary in size.
- *Site*—it is seen in mandible (**Fig. 16-20**) and are bilateral in occurrence. The lesion may be located in the periodontal ligament space.
- *Appearance*—an appearance that is highly suggestive of metastatic carcinoma is the combination of an area of bone destruction having very irregular margins with islands of bone within radiolucent areas.
- *Moth eaten appearance*—the other appearance of the lytic type of metastasis in the jaw is one that has considerable

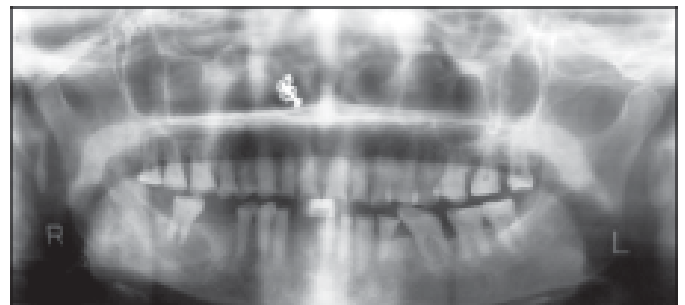


Fig. 16-20: Osteolytic variety of metastatic carcinoma showing radiolucency in mandibular right posterior region (Courtesy Dr Ashok L).

resemblance to some manifestation of osteomyelitis. There is an area of bone destruction or there may be two or more separated areas.

- *Infiltrative margin*—irregular bone destruction takes place from each separated site of origin and there is evidence of infiltrative margins in the areas of destruction.
- *Punched out radiolucency*—there may be multiple punched out radiolucency when several nests of tumor cells are located close to each other.
- *Margins*—it may be well demarcated but without cortication or encapsulation. They may have ill defined invasive margins.
- *Periodontal ligament widening*—in some metastatic tumor, there is widening of periodontal ligament.
- *Effect on surrounding structures*—if it occurs in alveolar bone, there is loss of lamina dura leading to mobility or loss of teeth. If the tumor has seeded in the papilla of a developing tooth, the cortices of crypts may be totally or partially destroyed. The cortical structures such as the neurovascular canal, sinus and nasal fossa are destroyed.

Osteoblastic type (Fig. 16-21)

- In some cases, osteoblastic variety of metastatic carcinoma can occur which depend upon whether the metastasized tumor cell produce significant level of acid or alkaline phosphatase.
- *Salt and pepper appearance*—irregular ‘salt and pepper appearance’ can be seen in some cases. Metastatic tumor widely disseminate in multiple nests in the bone. These nests appear as small radiolucent (pepper) areas. They induce sclerosis about themselves and thus separate the overall image with small radiopaque foci (salt).



Fig. 16-21: Osteoblastic variety of metastatic carcinoma
(Courtesy Dr Ashok L).

Diagnosis

- *Clinical diagnosis*—unexplained mobility of teeth, unhealed socket with numb chin syndrome may suspect metastatic carcinoma.
- *Radiological diagnosis*—salt and pepper appearance are seen in osteoblastic variety and ill defined radiolucency are seen in osteolytic variety.
- *Laboratory diagnosis*—biopsy features are similar to squamous cell carcinoma.

Differential Diagnosis

- *Exophytic squamous cell carcinoma*—there is history of primary tumor. Clinically squamous cell carcinoma is well distinguishable.
- *Multiple myeloma*—the borders of multiple myeloma are better circumscribed than metastatic tumor.
- *Periapical inflammatory lesion*—if the metastatic tumor is situated in the periodontal ligament space then it may be confused with metastatic tumor. But in case of inflammatory lesion, periodontal ligament space widening is more centered around the apex of the teeth and in metastatic tumor there is irregular widening.
- *Malignant salivary gland tumor*—it is not so common in mandible.

Management

- *Combination therapy*—It can be treated by chemotherapy, radiation therapy, surgery, immunotherapy and hormone therapy. Prognosis is poor and death occurs within a short time.

Basal Cell Carcinoma

It is also called as ‘*basal cell epithelioma*’ or ‘*Rodent ulcer*’. It arises from basal layer of epidermis or from the hair follicle.

Etiology

- *Prolonged exposure to sunlight*—the specific factor in sunlight responsible for skin carcinogenesis appears to be an ultraviolet radiation.
- *Others factors*—other factors are also responsible like burn scars and ionizing radiation. General atrophy associated with aging process; at least, predispose to develop the skin cancer.

Clinical Types

- *Nodular (noduloulcerative) basal cell carcinoma*—clinically, it appears as nodular lesion with central depression.
- *Pigmented basal cell carcinoma*—tumor is colonized by benign melanocytes.

- *Sclerosing (morpheaform) basal cell carcinoma*—it mimics scar tissue.
- *Superficial basal cell carcinoma*—occurs on skin of the trunk.
- *Nevoid basal cell carcinoma*—basal cell carcinoma is associated with syndrome.

Histological Types

- *Adenoid basal cell carcinoma*—it represent neoplasm which mimics glandular formation.
- *Cystic basal cell carcinoma*—presence of many cysts in the lesion.
- *Keratotic basal cell carcinoma*—formation of parakeratotic cells and horn cysts and attempted formation of hair structures to the trichoepithelioma.
- *Solid or primordial basal cell carcinoma*—cells have little tendency to differentiate.

Clinical Features

- *Age and sex distribution*—it is seen in middle aged or elderly person usually in 4th decade of life. It is much more common in men than women because men are exposed to the environmental elements more than women.
- *Site*—it develops most frequently on exposed surface of skin, middle-thirds of face and the scalp. There is also involvement of lip. The upper lip is involved more commonly than the lower lip.
- *Race*—blond people with fair complexion who have spent much of their lives outdoors are often victim of these lesions.
- *Appearance*—it begins as a small, slightly elevated papule which ulcerates, heals over and then breaks down again to form crusted ulcer.
- *Margins*—it develops a smooth, rolled border representing tumor cells spreading laterally beneath the skin (Fig. 16-22).



Fig. 16-22: Rolled margin seen in basal cell carcinoma showing spread in the skin.

- *Rodent ulcer*—untreated lesion continues to enlarge and infiltrate the adjacent and deeper tissues and it may even erode deeply into the cartilage or bone. Due to its invading and destructive infiltration into adjoining tissues, it gradually increases in size and accounts for its synonym 'rodent ulcer' (Fig. 16-23).



Fig. 16-23: Destruction of skin occur in basal cell carcinoma signifying its name as 'rodent ulcer'.

- *Intraoral lesion*—it is never seen in oral cavity unless it arrives there by invasion and infiltration from a skin surface.
- *Nodular (noduloulcerative) basal cell carcinoma*—it is firm painless papules with central depression and has umbilicated appearance. When the lesion is pressed, characteristic pearly opalescent quality is seen.
- *Pigmented basal cell carcinoma*—the melanin production imparts a tan, brown, black color of the lesion. In some cases, it is even bluish in color.
- *Sclerosing (morpheaform) basal cell carcinoma*—in this, overlying skin appear pale, atrophic and lesion is firm on palpation. Margins are poorly demarcated.
- *Superficial basal cell carcinoma*—it is multiple, well demarcated lesions seen on skin. It is also erythematous scaly patches some time mistaken as psoriasis.
- *Lymph node involvement*—occasionally, it may metastasize to lymph nodes.

Diagnosis

- *Clinical diagnosis*—rolled margin with ulcerative lesion on the skin of the face with destruction of skin, suggestive of basal cell carcinoma.
- *Laboratory diagnosis*—biopsy will show cell nests which are composed of a layer of cells, usually well polarized, that are strongly suggestive of the basal cell layer of skin.

Management

- *Simple surgical excision*—it is indicated in small lesion which is less than 1 cm in diameter. Surgery can be performed by laser ablation, electrodesiccation and curettage.
- *Radical surgical excision and radiotherapy*—it is given in extensive and aggressive lesion.
- *Mohs micrographic surgery*—it is indicated in sclerosing lesion. It is used for frozen section evaluation if any tissue is left behind. If tissue remains then surgeon remove the tissue immediately and process is repeat until patient is free of disease.

Transitional Cell Carcinoma

These lesions arise chiefly from the tonsil, base of the tongue and nasopharynx due to presence of stratified epithelium in this area. It is extremely malignant, running a rapid clinical course, metastasizing widely and causing very early death.

Clinical Features

- *Age and sex distribution*—mean age of occurrence is 44 years.
- *Symptoms*—there may be sore throat, nasal obstruction, defective hearing or ear pain, headache, dysphagia, epistaxis and ocular symptoms.
- *Primary lesion*—the primary lesions are very small often completely hidden, usually slightly elevated (**Fig. 16-24**). In some cases, primary lesion is ulcerated with granular eroded surface.
- *Advanced lesion*—as the tumor is advanced, its size is increased rapidly and base of tumor appear indurated. In some instances, it appears as an exophytic or fungating growth.
- *Lymph nodes*—swelling of regional lymph node is the most common occurrence.



Fig. 16-24: Carcinoma seen in tonsillar area occur due to tonsillar cell malignancy.

Diagnosis

- *Clinical diagnosis*—any ulcerative growth in the region of tonsil, nasopharynx and base of tongue will suspect transitional cell carcinoma.
- *Laboratory diagnosis*—biopsy shows cells growing in solid sheets or in cords and nests. The individual cell is moderately large, round or polyhedral and exhibit a lightly basophilic cytoplasm and indistinct cell outlines. There are large nuclei with presence of mitotic activity.

Management

- *Radiation*—X-ray radiation is the most commonly accepted treatment.

Malignant Melanoma

It is also called as '*melanosarcoma*'. It is a neoplasm of epidermal melanocytes. It is one of the biologically unpredictable and deadly of all human neoplasms.

Etiology

- *Sunlight*—it is very important etiological factor in cutaneous melanoma. Acute sun damage causing melanoma is more as compared to chronic sun damage.
- *Genetic*—chances of malignant melanoma increases in a person whose relative has got the history of cancer.
- *Other factors*—other factors like fair complexion, history of blistering sunburns, indoor occupation with outdoor recreation habits and history of dysplastic or congenital nevus may be contributed for the development of malignant melanoma.

Types

- *Superficial spreading melanoma*—It is the most common cutaneous melanoma.
- *Nodular melanoma*—it is a form of cutaneous melanoma and occurs in head and neck area.
- *Lentigo maligna melanoma*—it develops from precursor lesion called lentigo maligna.
- *Acral lentiginous melanoma*—it is the most common form of melanoma in black. It is most common in oral cavity.

Growth Pattern

- *Radial growth phase*—the radial growth phase may last for several months to several years. In this phase, malignant melanocytes spread horizontally through basal layer of epidermis.
- *Vertical growth phase*—the vertical growth phase is characterized by increase in size, change in color, nodularity and at times, ulceration. In this phase, malignant cells invade underlying connective tissue.

Clinical Features

Superficial spreading melanoma

- **Age**—The majority of cases are diagnosed in the 5th to 7th decade of life. It is more common in males as compared to females in the ratio of 2:1.
- **Site**—the lesion presents as a tan brown, black or admixed lesion on sun exposed skin, especially in blacks. It also occurs on skin of face (**Fig. 16-25**), head and neck, chest, abdomen and extremities.



Fig. 16-25: Malignant melanoma seen on face of patient showing ulceration.

- **Appearance**—it begins as pigmented macule in superficial radial growth pattern, restricted mostly to epithelium and junction. In advanced cases, melanoma present as an ulcerated, fungating growth which is associated with bleeding.
- **Amelanotic melanoma**—Some lesions are devoid of pigmentation which is called as amelanotic melanoma.

Nodular melanoma

- **Site**—they have a predilection for occurrence on head and neck of men.
- **Color**—color varies from mucosal pink through brown and blue to black.
- **Consistency**—it is firm on palpation.
- **Margins**—it has got erythematous borders which surround the tumor.
- **Extent**—there is rapid infiltration in nodular type of melanoma. It may be focal or diffuse.
- **Signs**—many times, it may ulcerate and hemorrhage may be seen. In later stages, it becomes more diffuse, nodular and tumefactive with foci of hypo- and hyperpigmentation. It presents as a sharply delineated nodule with some degree of pigmentation.

Lentigo maligna melanoma

- **Sex**—it occurs more often in women than in men.
- **Site**—it has got predilection for exposed parts.

- **Margins**—it is pigmented macule with ill defined margins.
- **Appearance**—it occurs characteristically as a macular lesion of the malar area.
- **Progress**—it grows slowly in radial phase, with around the central axis and in a superficial manner.
- **Extent**—as the disease advances, invasion and metastasis are frequent.

Acral lentiginous melanoma

- **Synonym**—it is also called as mucosal lentiginous melanoma.
- **Site**—it develops on palms of hands, soles of the feet, sublingual area and mucous membrane.
- **Appearance**—it develops as dark pigmented irregularly margined macule.

Oral Manifestations

- **Age and sex**—it is an uncommon neoplasm of the oral mucosa and it is more common in men than in women with the overall age of occurrence being 55 years, with most cases occurring between 40 to 70 years.
- **Site**—it has got definite predilection for palate and maxillary gingiva/alveolar ridge followed by buccal mucosa, mandibular mucosa, tongue, lips and floor of mouth.
- **Preceding factors**—focal pigmentation preceding the development of actual neoplasm frequently occurs, several months to years before clinical symptoms appear.
- **Symptoms**—oral melanomas are generally painless. The lesion presents as a soft, darkish brown or black mass. It may have a nodular or a papillary surface.
- **Appearance**—the lesions usually appear as deeply pigmented areas, at times ulcerated and hemorrhagic (**Fig. 16-26**), which tend to increase progressively in size.



Fig. 16-26: Intraoral malignant melanoma presented as darkly pigmented brownish lesion with ulceration.

- **Signs**—the tumor causes extensive destruction of the underlying bone; loosening of teeth may be a concomitant clinical finding. There may be ulcerations and bleeding of the oral mucosa.
- **Base and margins**—there is minimum induration and no rolled margins are seen.

Radiographic Features

- **Radiodensity**—some melanomas in the jaws may present radiographic picture which is indistinguishable from osteomyelitis. Sometimes, small radiolucency with irregular border may be seen (**Fig. 16-27**).

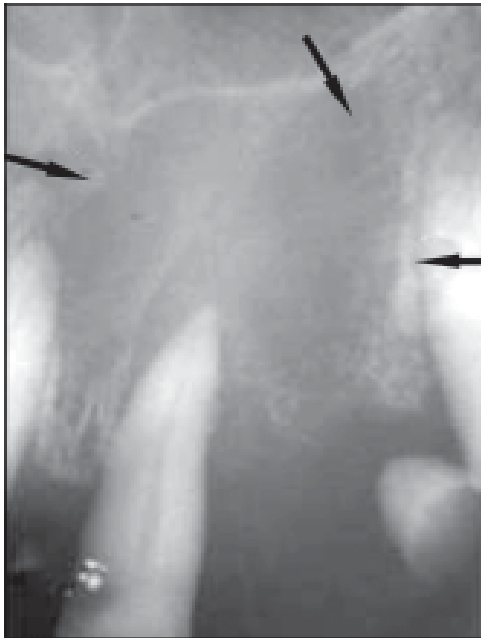


Fig. 16-27: Radiolucency seen in upper region which is irregular in shape in case of malignant melanoma (Courtesy Dr Parate).

Diagnosis

- **Clinical diagnosis**—darkly pigmented lesion with ulceration will give clue to the diagnosis.
- **Radiological diagnosis**—not specific.
- **Laboratory diagnosis**—in superficial spreading melanoma melanocytes are arranged in “pagetoid manner”. In case of nodular melanoma, there are large, epithelioid melanocytes within the connective tissue. Small ovoid and spindle shaped cells may be present. Lentigo melanoma is characterized by increased number of atypical melanocytes with the basal epithelial layer.

Histological Grading of Malignant Melanoma

- It depends upon the depth upto which malignant cells have invaded or infiltrated into the connective tissue.
- **Grade I**—malignant cells are confined within the epithelium.

- **Grade II**—malignant cells have invaded into the papillary dermis.
- **Grade III**—malignant cells have invaded into the level of reticular dermis.
- **Grade IV**—malignant cells have completely invaded the reticular dermis.
- **Grade V**—malignant cells have extended into the subcutaneous fat.

Management

- **Surgical excision**—it is treated by surgical excision. Surgical removal of regional lymph nodes should also be carried out.
- **Other therapy**—other therapy like irradiation, immunotherapy and chemotherapy or, by combination of these methods can also be tried. Survival rate is very poor and is worse with metastasis.

Verrucous Carcinoma

It is a slow growing low-grade carcinoma. It is also called as ‘Snuff dipper cancer’, ‘Ackerman’s tumor’. It is reported by Ackerman in 1948 as spit tobacco associated malignancy.

Etiology

- **Tobacco**—tobacco chewers have high percentage of these cases. It occurs usually in a person habitual to hold the quid in the buccal sulcus (**Fig. 16-28**).



Fig. 16-28: Verrucous carcinoma seen in buccal mucosal area where patient is used to keep tobacco (Courtesy Dr Bhaskar Patle).

Clinical Features

- **Age and sex**—it is generally seen in elder population with mean age of occurrences of 60 to 70 years. Men are affected more as compared to women.

- **Common sites**—they are larynx, external auditory meatus, lacrimal duct, skin, scrotum, penis, vulva, vagina, uterine cervix, perineum, leg and odontogenic cyst linings. In the oral cavity, it occurs on buccal mucosa (Fig. 16-28) and gingiva or alveolar ridge.
- **Symptoms**—pain and difficulty in mastication are common complaints.
- **Appearance**—they appear papillary in nature with pebbly surface which is sometimes covered by a white leukoplakic film (Fig. 16-29). They have rugae-like folds with deep cleft between them (Fig. 16-30). In some cases, there may be warty fungating mass.
- **Base**—large broad lesion with minimum to extensive elevation above the surface of mucosa.
- **Margins**—margins are well defined and show rim of slightly elevated normal mucosa (Fig. 16-31).
- **Color**—color is usually white but it can be erythematous or pink.



Fig. 16-29: Pebbly surface of verrucous carcinoma surrounded by leukoplakic film (Courtesy Dr Parate).



Fig. 16-30: Warty growth with cleft in between in verrucous carcinoma (Courtesy Dr Chole).



Fig. 16-31: Well defined and slightly elevated margins in verrucous carcinoma.

- **Lymph nodes**—regional lymph nodes are often tender and enlarged simulating metastatic tumor, but the node involvement is usually inflammatory.
- **Extent**—lesion on the mandibular mucosa grows into overlying soft tissue and rapidly becomes fixed to the periosteum, gradually invading and destroying the mandible.
- **Progress**—oral lesion is slow growing, chiefly exophytic and only superficially invasive, at least until late in the course of the disease and has a low metastatic potential.
- **Prognosis**—prognosis in verrucous carcinoma is very good because of absence or late appearance of metastases.

Diagnosis

- **Clinical diagnosis**—verrucous papillary surface with cleft in between with history of tobacco keeping at location where lesion is found.
- **Laboratory diagnosis**—in biopsy, there is marked epithelial proliferation, parakeratin plugging and pushing margin.

Differential Diagnosis

- **Verrucous hyperplasia**—verrucous hyperplasia is a proliferative epithelial lesion with epithelial hyperplastic fold extending above the margins of surrounding mucosa whereas in verrucous carcinoma, folds invade down into connective tissue and hence are below the surrounding normal mucosal margin.
- **Chronic hyperplastic candidiasis**—in it, epithelial hyperplasia is not so extensive as in verrucous carcinoma and the cleft-like spaces often seen between the heaped up masses of carcinomatous epithelium.
- **Well differentiated squamous cell carcinoma**—lymph nodes metastasis is extensive. Histopathological investigation should be carried out to confirm the diagnosis.

Management

- *Surgical excision*—excision must be sufficiently radical to remove the entire lesion.

Spindle Cell Carcinoma

It is also called as 'Lane tumor', 'polypoid squamous cell carcinoma' or 'carcinosarcoma'. It occurs chiefly in respiratory and alimentary tracts. It is a variant of squamous cell carcinoma. There is proliferation of spindle cells believed to be arising from the surface epithelium. Many cases of spindle cell carcinoma develop as a recurrence after radiotherapy to the squamous cell carcinoma. This phenomenon is called as 'dedifferentiation'.

Clinical Features

- *Sex and age distribution*—it is more common in male with mean age of occurrence of 57 years.
- *Common site*—lower lip, tongue and alveolar ridge or gingiva with remainder scattered at other site.
- *Symptoms*—there is swelling, pain and presence of a non-healing ulcer.
- *Appearance*—the initial lesions appear either with a polypoid, exophytic or endophytic configuration. The lesion is fleshy.
- *Progress*—it grows rapidly and diagnose at later stage.

Diagnosis

- *Clinical diagnosis*—rapidly growing polypoid exophytic mass can yield in the clinical diagnosis of the lesion.
- *Laboratory diagnosis*—biopsy will show proliferation and 'dropping off' of basal cell to spindle cell.

Management

- *Radical surgery*—surgical removal of tumor with or without radical neck dissection is usually recommended.
- *Radiotherapy*—radiation therapy is also used in some cases.

Adenoid Squamous Cell Carcinoma

It is also called as 'adenoacanthoma'. It arises from pilosebaceous structures or senile keratosis with acantholysis.

Clinical Features

- *Sex and age distribution*—females are affected more with age ranging from 20 to 50 years and older.
- *Common site*—it more commonly occurs on lip and also in head and neck region. The lower lip is affected more commonly than upper lip and common on the vermillion border of lip.

- *Appearance*—it appears as simply elevated nodules (Fig. 16-32) that may be slow crusting, scaling. Usually, there is no surface ulceration.
- *Margins*—sometimes, there are elevated or rolled borders of the lesion.
- *Prognosis*—prognosis is good and metastasis is rare.

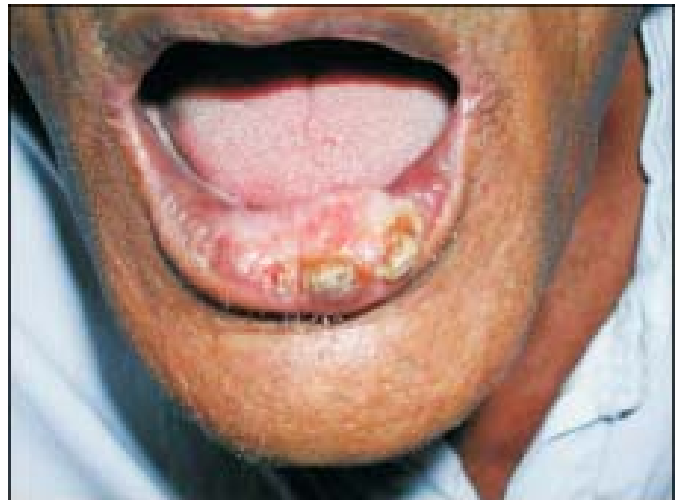


Fig. 16-32: Nodular elevated lesion seen on lower lip suggestive of adenoid squamous cell carcinoma (Courtesy Dr Suwas Darvekar).

Diagnosis

- *Clinical diagnosis*—lesion on lower lip with nodular surface and scaling will yield in diagnosis.
- *Laboratory diagnosis*—in biopsy, epithelium show the characteristic solid and tubular ductal structures which are lined by a layer of cuboidal cells and often contain or enclose acantholytic or dyskeratotic cells.

Management

- *Surgical excision*—it is treated by surgical excision.

Nasopharyngeal Carcinoma

It is tumor which arises from lining epithelium of lymphoid tissue rich nasopharynx. It can be caused by environmental factors, infection with Epstein-Barr virus, vitamin C deficient diet, and consumption of salt fish which contains carcinogen N-nitrosamines.

Clinical Features

- *Age and sex distribution*—it is common in older age group and more common in men as compare to women.
- *Onset*—the initial lesion is small and difficult to detect.
- *Symptoms*—patient may notice serous otitis media, otalgia and hearing loss from obstruction of eustachian tube. Other symptoms noticed are nasal obstruction and pharyngeal pain. If the tumor involves brain, it may cause neurological symptoms.

- *Signs*—first sign is, firm to hard cervical lymph nodes which may be enlarged in size.

Diagnosis

- *Clinical diagnosis*—ear symptoms with pharyngeal pain with cervical lymphadenopathy may give clue to the diagnosis.
- *Laboratory diagnosis*—biopsy may show features suggestive of squamous cell carcinoma, differentiate non-keratinizing carcinoma, and undifferentiated non-keratinizing carcinoma.

Management

- *Radiotherapy*—as the tumor is inaccessible, it is more frequently treated with radiotherapy.

Multicentric Oral Carcinoma

Multiple primary tumors occur in about 10% of patients with oral cancer. It is usually seen in alcoholics and heavy smokers. The reason for this is that the whole mucosa is often in an abnormal state for long periods, prior to the development of oral cancer. The commonest finding is that there is area of atrophy, either adjacent to a carcinoma or randomly distributed.

Tumors of Fibrous Connective Tissues

Fibrosarcoma

It is malignant neoplasm composed of malignant fibroblast that produces collagen and elastin. It arises in the periosteal tissue. It may arise secondarily in tissues that have received therapeutic levels of radiation. It is one of the most common soft tissue sarcoma.

Clinical Features

- *Age and sex*—onset at any age with mean age of occurrence of 50 years with male predominance.
- *Common sites*—it is commonly associated with cheek, maxillary sinus, pharynx, palate, lip and periosteum of maxilla and mandible. Mandible is more commonly affected than maxilla.
- *Symptoms*—It produces fleshy bulky mass of tissue. There may be pain and loosening of the teeth. Involvement of TMJ and para-mandibular musculature are producing trismus. Sensory neural abnormalities may occur if it involves peripheral nerves.
- *Signs*—initially they resemble benign fibrous outgrowth, but they grow rapidly to produce large tumor. Large tumors are prone to ulceration and hemorrhage (Figs

16-33A and B). Secondary infections are seen in some cases. In some cases, pathological fracture may occur.

- *Maxillary lesions*—maxillary lesions are quite destructive and invade the antrum.
- *Extent*—they tend to penetrate the cortex and spread along the periosteum.



Figs 16-33A and B: Extraoral and intraoral view of fibrosarcoma showing extensive ulcerative growth.

Radiographic Features

- *Radiodensity*—destructive lesion may stimulate the osteolytic form of osteosarcoma.
- *Margins*—they are poorly demarcated, non-corticated and lack of any resemblance of capsule.
- *Extent*—they tend to elongate through marrow space.
- *Appearance*—if soft tissue lesions occur adjacent to bone, they cause a saucer-like depression (Fig. 16-34) in the underlying bone or invade it as seen in a squamous cell carcinoma.
- *Teeth*—teeth are displaced and loose their supporting bone so that they appear to be floating in space. Lamina dura and follicular cortices are obliterated. In some cases, periodontal space widening may occur.

- *Maxillary sinus*—inferior border of maxillary sinus, posterior wall of maxilla, nasal floor, inferior border of mandible and neurovascular canal are lost.
- *Pressure effect*—if it is developed from the periosteum, it may cause a smooth pressure resorption of underlying bone.

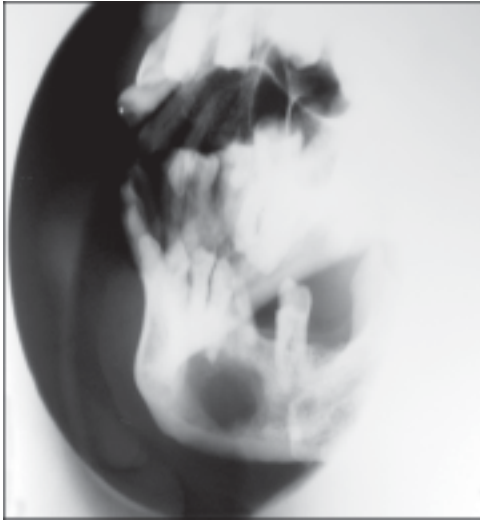


Fig. 16-34: Saucer shaped erosion seen in premolar area in fibrosarcoma.

Diagnosis

- *Clinical diagnosis*—fleshy mass with ulceration in the soft tissue area will suspect fibrosarcoma.
- *Radiological diagnosis*—saucer shaped erosion of bone is present.
- *Laboratory diagnosis*—proliferation of fibroblasts and formation of collagen and reticular fibers. Mitotic figures are prominent in small group of poorly differentiated tumors.

Differential Diagnosis

- *Metastatic carcinoma*—there is history of primary tumor and it usually does not cause enlargement of jaw.
- *Osteosarcoma*—there is typical sunray appearance.
- *Liposarcoma*—it is very rare in jaws.
- *Neurogenic sarcoma*—it is difficult to differentiate.

Management

- *Radical surgical excision*—it is most commonly used treatment modality.

Malignant Fibrous Histiocytoma

It is also called as '*malignant fibroxanthoma*'. Neoplasm that exhibits both histiocytic and fibrocytic features are referred to as histiocytomas. It is one of the most common soft tissue sarcoma of soft tissues of body.

Types (Histological)

- Giant cell
- Inflammatory
- Myxoid
- Storiform
- Pleomorphic
- Angiomatoid

Clinical Features

- *Age and sex*—there is slight male predilection and malignant variety is common in adults.
- *Site*—in the head and neck area, more commonly encountered in the paranasal sinuses or centrally, within the jaw.
- *Appearance*—occasionally, it arises in the oral cavity and in the lateral neck, it appears as indurated swelling.
- *Metastasis*—in nearly 1/4th of cases have been reported.

Radiological Features

- *Appearance*—it produces unilocular or multilocular radiolucent areas.
- *Margins*—lesion has got ill-defined borders.

Diagnosis

- *Clinical diagnosis*—not specific.
- *Radiological diagnosis*—not specific.
- *Laboratory diagnosis*—the spindle cells are arranged in fascicles with a pinwheel or storiform pattern. The deep seated tumors contain both epithelioid histiocytes and spindle shaped fibroblast. Both cell component display marked hyperchromatism, pleomorphism and atypical mitoses with many multinucleated cells exhibiting angulated cytoplasmic borders.

Management

- *Surgical excision*—it is treated by surgical excision or radiation with 5 year survival rate in 20 to 60% of cases.

Synovial Sarcoma

It usually arises from articular or para-articular sites, bursae or tendon sheath. Synovial sarcoma of head and neck are rare.

Clinical Features

- *Age and sex distribution*—it is predominately seen in young adults mean age being 19 years. It has got slight male predilection.
- *Sites*—intraoral sites are cheek, tongue, floor of mouth and soft palate.
- *Symptoms*—there is painless deep seated swelling which may produce difficulties in breathing or swallowing.

Diagnosis

- *Clinical diagnosis*—it is not specific.
- *Laboratory diagnosis*—it is characterized by biphasic cellular pattern of cleft-like or slit-like spaces lined by cuboidal epithelial-like cells. The space may contain PAS positive mucoid material. There may be fibrosarcoma like proliferation of cells, with associated collagen or reticulum.

Management

- *Radical resection*—early radical resection is the best method of treatment.
- *Adjunct radiotherapy*—it can also be given after radical resection.
- *Prognosis*—it has got poor prognosis as tumor often metastasized.

Tumors of Adipose Tissues

Liposarcoma

It is extremely uncommon malignant tumor of head and neck region.

Clinical Features

- *Age and sex*—it most frequently occurs in adults over the age of 40 years with predilection in males in ratio of 2:1.
- *Site*—the most common site is thigh, retroperitoneum and inguinal region. The most frequent oral region affected is cheek which is followed by tongue.
- *Symptoms*—pain and tenderness may be present in advanced cases of liposarcoma.
- *Appearance*—it has a slow, silent growth, submucosal or deep in location, producing firm, resilient lesions, sometimes lobulated.
- *Color*—color of lesion is normal or yellow in color.

Diagnosis

- *Clinical diagnosis*—soft, yellow color growth on the cheek may suspect liposarcoma.
- *Laboratory diagnosis*—histologically, it is classified into myxoid, round cell, adult and pleomorphic types. In general, it consists of fat cells and lipoblast in varying degrees of differentiation and anaplasia with variable stromal component.

Management

- *Surgical excision*—it is treated by surgical excision with or without radiation therapy.

Cartilage Tumors

Chondrosarcoma

It is also called as '*chondrogenic sarcoma*'. It develop from natural cartilage or a benign cartilaginous tumor. Most of them develop from cartilage located in bone either centrally in bone (medullary cavity) or peripherally, from the cartilage cup of an osteochondroma.

Types

- *Primary*—they directly arise from the cartilage.
- *Secondary*—it develops in a preexisting benign cartilaginous tumor.

Clinical Features

- *Age and sex*—develop during 3rd to 6th decades and male to female ratio is 2:1. Secondary chondrosarcoma occur at early age than the primary type of chondrosarcoma.
- *Site*—it is rare in jaws and may occur in maxilla or mandible. Often found in anterior alveolar process of maxilla and in mandible, it is found at angle and alveolar ridge of premolar-molar region.
- *Progress*—it is slow growing and less malignant.
- *Symptoms*—painless in early stages with facial asymmetry as first complain but as it enlarges, swelling becomes bony hard and painful. There may be headache.
- *Signs*—teeth adjacent to the lesion are resorbed, loosened and get exfoliated. In some cases, there may be hemorrhage from the neck of teeth. There may be sensory nerve deficit, proptosis and visual disturbances. If swelling erodes through the cortical plate, it tends to be tender, smoothly contoured firm mass due to presence of cartilage.
- *Surface of lesion*—mucosal covering appears normal in early stage (**Fig. 16-35**) but later, it ulcerates and develops necrotic surface, if chronically traumatized.
- *Temporomandibular joint involvement*—if it occurs in/or near the temporomandibular joint region (**Fig. 16-36**), trismus and abnormal joint function may result.
- *Metastasis*—metastatic spread by vascular channel. Malignant cells may erode through wall or venules and extend along inside the venules without adhering to vessels wall but still altered at their site of entry. Lung is common region of metastasis.

Radiographic Features

- *Radiodensity*—it may be sclerotic or mixed, if there is calcification of neoplastic tissue.
- *Margins*—lytic lesions with poorly defined borders.
- *Shape*—the lesion is round, ovoid or lobulated.



Fig. 16-35: Chondrosarcoma of palate showing displacement of teeth and normal appearing mucosa in early stage.



Fig. 16-36: Chondrosarcoma of condyle (Note the swelling in condylar region).

- *Flocculent or snow-like appearance*—it may be multi-loculated or develop as multiple radiolucency containing radiopaque foci. The center radiopaque structure has been described as 'flocculent', implying *snow-like* features.
- *Soap bubble appearance*—large lobules of cartilage may give a radiolucent soap bubble appearance.
- *Ground glass or granular appearance*—diffuse calcifications may be superimposed on a bony background that resembles granular or ground glass appearance.
- *Sunray appearance*—occasionally, peripheral periosteal new bone may be present perpendicular to the original

cortex, giving so called sunray appearances. Sometimes, ground glass appearance may be seen.

- *Speckled appearance*—careful examination of areas of flocculence may reveal a central radiolucent nidus which is probably cartilage, surrounded by calcification. The result is rounded or speckled areas of calcification.
- *Teeth*—there may be band-like widening of periodontal ligament space; resorption of root may occur.
- *Mandibular lesion*—in mandible, inferior border or alveolar process may be grossly expanded, while still maintaining its cortical covering.
- *Maxillary lesion*—maxillary lesion may push the walls of maxillary sinus or nasal fossa and impinge on the infratemporal fossa.
- *Condylar lesion*—lesion on the condyle cause it's expansion and remodeling of the corresponding articular fossa and eminence (**Fig. 16-37**). If the lesion occurs in the articular disc region, a widened joint space may be present with corresponding remodeling of condylar neck.
- *3D CT features*—it may demonstrate lesion very clearly (**Fig. 16-38**).

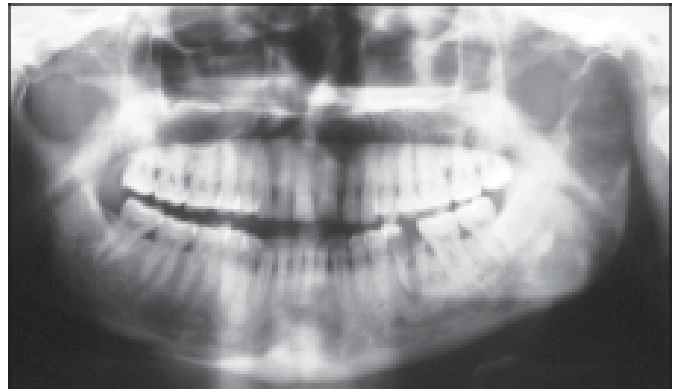


Fig. 16-37: Destruction of condyle on left side of patient in chondrosarcoma of condyle.

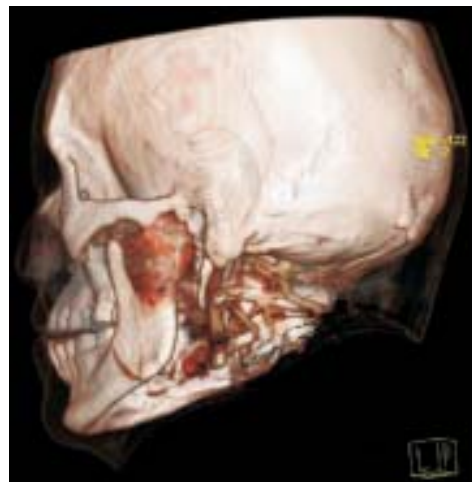


Fig. 16-38: Chondrosarcoma of condyle a 3D CT picture (Courtesy Dr Iswar).

Diagnosis

- *Clinical diagnosis*—this can be included in differential diagnosis of swelling of condylar region.
- *Radiological features*—flocculent appearance, sunray appearance and in some cases, ground glass appearance is seen.
- *Laboratory diagnosis*—biopsy shows hyaline cartilage, increased number of cells with plump nuclei. Histopathologically, it can be grade I, Grade II or Grade III chondrosarcoma.

Differential Diagnosis

- *Osteosarcoma*—the typical calcification of chondrosarcoma is not present in osteosarcoma.
- *Fibrous dysplasia*—the periphery of fibrous dysplasia is better defined. In fibrous dysplasia there is thinning of periodontal ligament space and in chondrosarcoma, there is usually a widening of periodontal ligament space.

Management

- *Surgery*—it is the only treatment of choice and there is 5 years survival rate. Complete resection should always be done to avoid recurrence.

Mesenchymal Chondrosarcoma

This is aggressive form of chondrosarcoma showing biphasic histopathological pattern. It contains mesenchymal cells. It tends to metastasize to unusual locations and that too after long period of time. Most of the tumors arise in bone but an appreciable number occur in soft tissues.

Clinical Features

- *Age*—it is seen in younger age group than usual type of chondrosarcoma. It is usually seen between the ages of 10 and 30 years.
- *Site*—it is most commonly seen in maxilla, skull bone, mandible and ribs. It also occurs in tubular bones and pelvis.
- *Symptoms*—there may be pain due to compression of nerve. In some cases, there may be swelling.
- *Sign*—pathological fracture may also occur.

Radiological Features

- They are same as chondrosarcoma

Diagnosis

- *Clinical diagnosis*—younger age group with complaints of pain and swelling in maxillary area.

- *Radiological features*—same as that of chondrosarcoma.
- *Laboratory diagnosis*—the mesenchymal chondrosarcoma consist of sheets of small, round or ovoid, undifferentiated cells interspersed by small islands of well differentiated cartilage which often show calcifications and metaplastic bone formation.

Management

- *Surgery*—surgical excision with wide margin is the treatment of choice. The prognosis is poor and most patients may die due to metastasis.

Bone Tumors

Osteosarcoma

It is also called as 'osteogenic sarcoma'. It is the most common malignant tumor originated within the bone. It is derived from osteoblasts in which tumor cells contain high level of alkaline phosphatase.

Etiology

- *After radiation therapy*—there are increased incidences of osteosarcoma in bone that has been irradiated.
- *Traumatic irritation*—it may be causative factor for osteosarcoma.
- *Fibro-osseous diseases*—osteogenic sarcoma may be seen in fibrous dysplasia and Paget's disease.
- *Others*—other causes which can cause osteosarcoma are genetic mutation and some viral causes.

Clinical Features

- *Age and sex*—mean age of occurrence in the jaws is 33 years and it is more common in males.
- *Site*—it is more common in long bones like femur and tibia and in jaw bone.
- *Progress*—it grows rapidly with a doubling time of 32 days and shows recurrence and early metastasis via bloodstream to lungs (Fig. 16-39).
- *Signs*—there is exophthalmos, blindness, nasal obstruction and epistaxis.

Oral Manifestations

- *Incidence*—it is rare and accounts for 7% of all osteosarcomas.
- *Site*—it is equal in maxilla and mandible. In the mandible, lesion is seen in body and in maxilla it occurs in antrum or alveolar ridge.
- *Symptoms*—swelling of a short history and is accompanied by pain. Affected tooth may become displaced or loose. Numbness of lip and chin may be



Fig. 16-39: Rapidly growing swelling seen in floor of mouth in osteosarcoma patient.

due to involvement of inferior alveolar nerve. There may be trismus and hemorrhage.

- *Sign*—there may be history of tooth extraction with nodular or polypoid reddish granulomatous appearing mass growing from tooth sockets. Expansion is very firm due to dense fibrous tissue which is produced. Initially, swelling is smoothly contoured and covered by normal mucosa (Fig. 16-40). When expansion becomes chronically traumatized, mucositis develops on surface. Later, the surface gets ulcerated and looks like whitish gray in color.



Fig. 16-40: Smoothly contoured swelling seen in maxillary anterior region in osteosarcoma patient (Courtesy Dr Pravin Lambade).

Radiographic Features

- *Periodontal ligament space widening*—there is symmetrical widening of periodontal ligament space in early stage of the disease.
- *Types*—it may develop into a frankly osteolytic stage, mixed stage, i.e. those which produce some bone and thirdly those which are almost entirely bone forming.

- *Frankly osteolytic*
 - *Margins*—lesions are unicentric and borders of the lesion are ill defined.
 - *Appearance*—there is moth eaten appearance.
 - *Cortical plate*—perforation and expansion of cortical margins by extension into sub-periosteal bone.
 - *Neurovascular canal*—mandibular lesion may destroy the cortex of neurovascular bundles. Alternatively, neurovascular canal may be symmetrically widened or enlarged.
 - *Lamina dura*—adjacent lamina dura may be destroyed.
 - *Pathological fracture*—pathologic fracture occurs more readily as in sarcoma extending more deeply into the bone.
 - *Maxillary sinus*—sarcomas which develop near the floor of maxillary antrum or nasal fossa result in destruction of bone with no demarcation on tumor from the air cavity.
 - *Spiking resorption of root*—there is resorption of root which results in tapered narrowing of the root.
- *Mixed (Fig. 16-41)*
 - The presence of new bone laid down deep to the periosteum may be slight or it may be gross and without any structures.
 - *Margins*—sarcomas with small amount of new bone formation usually present margins which are not well defined.
 - *Appearance*—there is evidence of bone formation as well as destruction. In some cases, there is a well defined area of bone destruction, with partially corticated borders and some bone within the tumor itself.
 - *Honeycomb appearance*—the bone within the radio-lucent area of destruction may take the form of strands, which may be few and intersecting, or may produce a more or less honeycomb appearance.

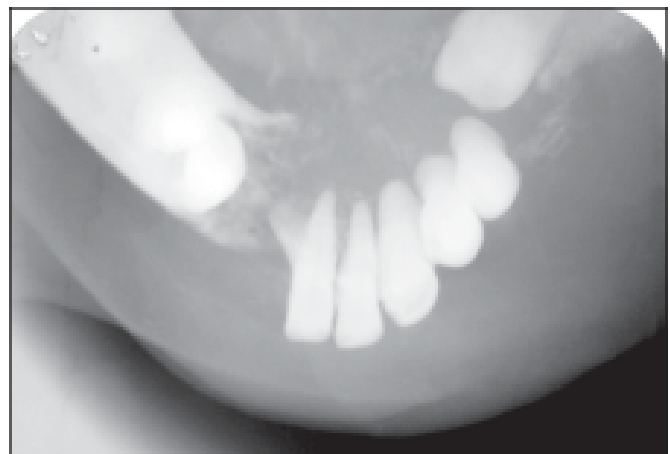


Fig. 16-41: Mixed type appearance seen in osteosarcoma patient. It also shows displacement and resorption of root.

- *Maxillary sinus and nasal fossa*—if the antrum or nasal fossae are involved, they are invaginated rather than infiltrated, since there is a complete bony covering over the tumor.
- *Frankly osteoblastic (sclerotic osteogenic sarcoma) (Fig. 16-42)*
- *Margins*—mixed lesion has ragged, ill defined borders and its radiographic pattern is the result of excessive bone production intermingled with radiolucent foci of bone destruction.

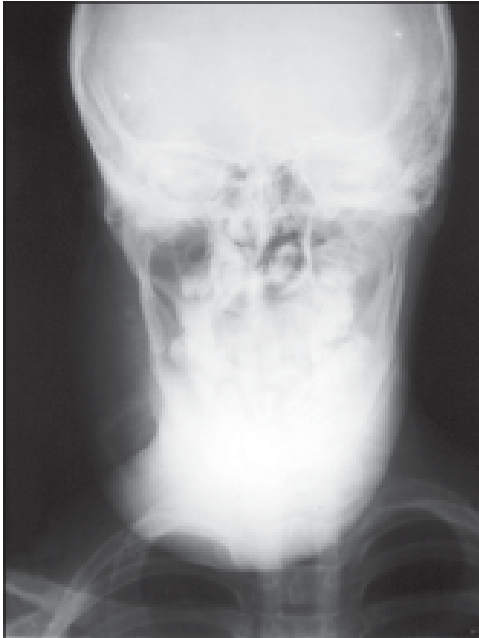


Fig. 16-42: Osteoblastic variety of osteogenic sarcoma showing radiopacity.

- *Granular appearance*—the sclerotic portion of mixed and opaque lesion may show vertical obliteration of trabecular pattern by new bone, impairing dense granular or sclerotic appearance.
- *Sunray appearance*—if the tumor has invaded the periosteum, many thin irregular spicules of new bone directed outwards and perpendicular to surface of the lesion are seen producing a 'sunray' appearance (Fig. 16-43).
- *Codman's triangle*—sometimes two triangular radiopacities project from the cortex and mark the lateral extremities of the lesion referred as 'Codman's triangle'. It is seen where the tumor breaks through the surface of the bone and it is the result of separation of the periosteum from the surface of the adjacent bone and subsequent ossification in the space formed between periosteum and bony surface.
- *Onion peel appearance*—on rare occasion, subperiosteal bone laid down in layers and it may take form of onion peel lamination.
- *Alveolar ridge*—there is also distortion of the alveolar ridge.



Fig. 16-43: Typical sunray appearance seen in osteogenic sarcoma.

Variant of Osteosarcoma

- *Peripheral (Juxtacortical)*
 - *Parosteal osteosarcoma*—it is attached to the cortex by short stalk. In this, there is no elevation of periosteum and no peripheral reaction. It is uncommon, extremely rare in the jaws and is characterized by slow growth. It has good prognosis due to low tendency of tumor for metastasis. It is treated by radical excision which results in low risk of recurrence.
 - *Periosteal osteosarcoma*—it arises within the cortex and elevate the overlying periosteum. It is an aggressive variant of parosteal type of osteosarcoma. There is occurrence of periosteal new bone formation. This may perforate the surface of periosteum and lesion may extend into surrounding soft tissue. In this, radical surgical excision with wide margin is the treatment of choice.
- *Extra-osseous*—osteosarcoma of soft tissue in absence of primary skeletal tumor and occur in breast, liver and kidney. It is highly malignant type of tumor.
- *Post-irradiation bone sarcoma*—it can develop after 3 years of radiation. It is dose related. If higher dose is given, there are more chances of sarcoma.

Diagnosis

- *Clinical diagnosis*—rapidly growing swelling of bone which is accompanied by pain and discomfort will give clue to diagnosis.
- *Radiological diagnosis*—periodontal ligament space widening, sunray appearance, Codman's triangles are typical radiological features of the osteosarcoma.
- *Laboratory diagnosis*—serum alkaline phosphatase level is increased in osteosarcoma. Biopsy shows atypical osteoblasts. Neoplastic osteoblasts are spindle shaped or polyhedral. Pleomorphism in size and shape of cells.

Differential Diagnosis

- **Carcinoma**—in frank osteolytic stage, this can be confused; but, carcinoma arising from oral mucosa has a tendency to extend along the surface of the bone more than sarcoma itself which originated within the bone. The absence of any soft tissue involvement may suggest its central nature. Sarcoma is expected to extend more deeply into the bone with more frequent pathologic fracture. Complete dissolution of continuity of the mandible may take place more quickly with sarcoma than carcinoma. Sarcoma is more likely to produce bone destruction in a more concentric manner since it starts in the substance of bone, instead on the surface.
- **Peripheral fibroma with calcification**—benign, slow growth.
- **Ossifying subperiosteal hematoma**—history of recent trauma.
- **Chondrosarcoma**—it usually occurs in older age and in maxilla.
- **Fibrous dysplasia**—these are well demarcated than osteosarcoma and have more uniform internal structures.
- **Osteomyelitis**—osteosarcoma is not associated with signs of infection.

Management

- **Surgery**—radical resection with amputation of bone can be carried out.
- **Adjuvant chemotherapy**—sometimes, adjuvant chemotherapy can be instituted.

Ewing's Sarcoma

It was first described in 1921 by James Ewing. It is also called as 'round cell sarcoma' or 'endothelial myeloma'. It is derived from mesenchymal connective tissue of bone marrow.

Origin

- **Endothelial element**—it may arise from endothelial elements in the marrow.
- **Neuroblastoma**—it may be a secondary deposit of neuroblastoma.
- **Marrow spaces**—it may arise from reticulum cell lining of the marrow spaces.
- **Metastatic tumor**—some considered it to be a metastatic tumor, the primary of which is located in different sites, including bronchus.

Clinical Features

- **Age and sex**—it occurs between the ages of 5 and 25 years with a male to female ratio of 2:1.

- **Site**—the bones affected are the long bone of extremities although the skull, clavicle, ribs, shoulder and pelvic girdles are involved. 10 to 15% occur in jaw, usually in mandible.
- **Progress**—it is a very rapidly growing highly invasive tumor with early and widespread metastasis.
- **Symptoms**—initially, intermittent pain which later becomes continuous and is associated with rapid growth of the tumor and enlargement of bone. During the attacks of pain the tumor enlarges visibly. Pain is associated with febrile attacks and leukocytosis.
- **Metastasis**—it usually metastasizes to other bones.
- **Signs**—the swelling is hard but occasionally, it may be soft and fluctuant. In the early lesion, when the tumor is intraosseous, swelling is firm. When the tumor breaks through the cortex, it spreads extensively in the soft tissues and form a soft mass which may ulcerate. Swelling is warm and tender.
- **Surface**—there may be hyperemia of the overlying tissues (**Fig. 16-44**), suggesting inflammatory condition.
- **Other features**—facial neuralgias, lip paresthesia are present. Teeth may become mobile and paresthesia may develop. There may be trismus, epistaxis, exophthalmos and sinusitis.



Fig. 16-44: Extraoral swelling showing hyperemia of overlying tissue of Ewing's sarcoma.

Radiographic Features

- **Radiodensity**—it is ill-defined destructive radiolucent, lesion which may be unilocular or multilocular (**Fig. 16-45**).
- **Site**—most commonly seen in mandibular posterior area.
- **Margins**—it has ill-defined margins and is never corticated. In advanced cases, bone is destroyed in

uneven fashion, resulting in ragged borders. Areas of sclerosis may be found around the margins of the lesion with mottled rarefaction.

- *Onion skin appearance*—if it penetrates the cortex, it may stimulate the periosteum to produce thin layers of bone which are laid down in layers more or less, parallel to shaft: resulting in laminated or onion skin effect along the bone surface.
- *Sunray appearance*—in advanced cases, where osteophytes formation is present, it may present as sunray appearance.
- *Effect on surrounding structures*—it may cause pathologic fracture. There may be destruction of lamina dura and the supporting bone of adjacent teeth. Mandibular neurovascular canal, inferior border of mandible and alveolar cortical plate may be destroyed.

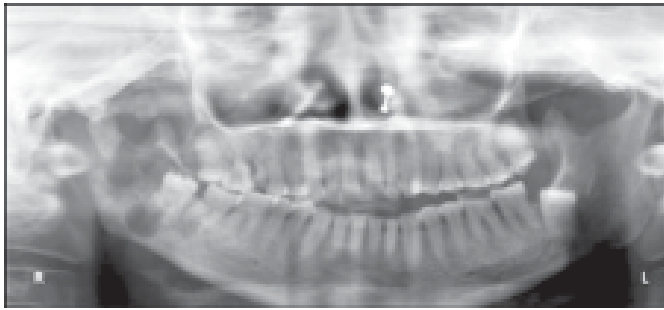


Fig. 16-45: Radiolucent lesion seen in ramus area in Ewing sarcoma.

Diagnosis

- *Clinical diagnosis*—intermittent pain in the bone with rapidly growing swelling may suspect this lesion.
- *Radiological diagnosis*—onion skin appearance is seen.
- *Laboratory diagnosis*—biopsy shows cellular neoplasm which is composed of solid sheets or masses of small round cells with very little stroma, although few connective tissue septa can be seen.

Differential Diagnosis

- *Osteomyelitis*—in osteomyelitis, there is presence of sequestra within the confines of lesion.
- *Eosinophilic granuloma*—it is associated with laminar periosteal bone reaction whereas Ewing's sarcoma does not produce laminar bone reaction.
- *Osteosarcoma, chondrosarcoma*—these are difficult to differentiate radiologically from Ewing's sarcoma. In it, histopathological investigation is conclusive.

Management

- *Combination therapy*—surgery combined with multidrug chemotherapy gives more survival rate nowadays.

Vascular Tumors

Malignant Hemangioendothelioma

It is a neoplasm of mesenchymal origin which is angiomatous in origin and derived from the endothelial cells.

Clinical Features

- *Age and sex*—it has slight predilection for females. It can arise at any age and has been found at birth also.
- *Site*—it may occur anywhere in the body but most commonly found in skin and subcutaneous tissues. In oral cavity, it can occur on lips, palate, gingiva, tongue and centrally within the maxilla and mandible.
- *Symptoms*—localized swelling with pain may be the feature of lesion.
- *Appearance*—it appears as flat or slightly raised lesion of varying size, dark red or bluish red in color.
- *Sign*—lesion can be ulcerated and show a tendency to bleed even after slight trauma.
- *Bone destruction*—bone may be involved by tumor producing a destructive process.

Diagnosis

- *Clinical diagnosis*—not so specific.
- *Laboratory diagnosis*—biopsy shows endothelial cells arranged in columns. Capillary formation is poorly defined, although anastomosing vascular channels may be discerned. The individual cells are pleomorphic large polyhedral or slightly flattened, with a faint outline and a round nucleus with multiple minute nucleoli.

Management

- *Surgery*—surgical excision of the lesion should be carried out.
- *Radiation*—surgery and X-ray radiation can be given.

Angiosarcoma

Angiosarcomas are malignant vascular tumors that comprise only 2% of soft tissue sarcoma. It is very rare in all sites including oral cavity.

Clinical Features

- *Site*—on the head it affect scalp. In the oral cavity, it appears in lip, palate, tongue, floor of mouth, cheek and gingivae. Mandible is more commonly affected than maxilla.
- *Appearance*—it appears as rapidly growing lesion that tends to ulcerate.

- *Sign*—margins of the lesion are ill defined. Surface is firm on palpation. There may be spontaneous bleeding seen from the lesion.
- *Lymph nodes*—regional lymph nodes are usually enlarged.

Radiological Features

- *Conventional film*—ill-defined ragged destruction of the bone occurs.
- *CT scan*—it reveals extension of soft tissue.

Diagnosis

- *Clinical diagnosis*—no specific.
- *Radiological diagnosis*—ill-defined destruction of bone can occur.
- *Laboratory diagnosis*—there are irregular vascular channels lined by endothelial cells that are often pleomorphic and may show numerous mitoses.

Management

- It is done by combination of surgery and radiation therapy.

Tumors of Neural Tissues

Neuroblastoma

It is a rare malignant tumor of the adrenal gland. It sometimes arises in the nerves or ganglia of the sympathetic system in the neck, thorax or abdomen. After leukemia, it is the commonest malignancy of children.

Clinical Features

- *Age*—it usually occurs in children under the age of 5 years.
- *Site*—oral cavity and nasal sinuses are involved. Oral involvement is usually due to metastasis and it occurs in mandible and maxilla.
- *Symptoms*—there is swelling, displacement of teeth and neurological symptoms.
- *Metastasis*—skull is one of the commonest sites for metastases. Metastases in bone tend to be multiple.

Radiological Features

- *Radiodensity*—some cases reveal areas of bone destruction with evidence of new bone formation within them and beneath the periosteum.
- *Sunburst appearance*—in some cases, there are spicules of bone within the soft tissue mass associated with bone destruction, these are roughly perpendicular to the surface of the bone and may resemble the 'sunburst' appearance.

- *Spoke wheel appearance*—in some cases, the spicules of bone or trabeculae are arranged radially in a rough architecture of wheel or spokes. The margins of the area of bone destruction are irregular and do not present any cortication.
- *Mottle area of decalcification*—a mottled area of decalcification is seen in some of the cases of metastases and with the passage of time, the lesion tends to become obviously osteolytic. Such areas may be localized or diffuse.

Diagnosis

- *Clinical diagnosis*—if neurological symptoms with swelling in jaw is present then neuroblastoma should be suspected.
- *Radiological diagnosis*—spoke wheel pattern, sunburst appearance are present.
- *Laboratory diagnosis*—biopsy shows degree of differentiation. It consists of small round cells arranged merely in diffuse masses. In some cases, differentiating cells begin to form groupings; generally referred to as rosettes and the cell themselves develop neurofibrillary processes.

Management

- *Surgery and radiation*—surgery and radiation is the treatment of choice in neuroblastoma.

Neurofibrosarcoma

It is also called as '*malignant Schwannoma*', '*malignant peripheral nerve sheath tumor*', '*neurogenic sarcoma*'. It arises from nerve tissue, especially from nerve sheath cells. It is rare in oral cavity.

Peripheral nerves are nerves that receive messages from the central nervous system (brain and spinal cord) leading them to stimulate voluntary movement. Neurofibrosarcoma, also known as peripheral nerve sheath tumor, is a malignant tumor that develops in the cells surrounding these peripheral nerves. It can sometimes arise in patients with neurofibromatosis.

Clinical Features

- *Sex and age distribution*—It usually occurs in 3rd and 6th decade with no sex predilection.
- *Common sites*—it occurs on lips, gingiva, palate and buccal mucosa. In central tumor mandible is more affected than maxilla.
- *Symptoms*—only complaint is the presence of mass and in some cases, pain or paresthesia may be present.
- *Appearance*—tumor mass sometimes exhibits rapid growth in patient.

Radiological Features

- **Radiodensity**—it reveals diffuse radiolucency is the characteristic feature of a malignant infiltrating neoplasm.
- **Appearance**—in some cases, it may appear as a smooth radiolucency. When the tumor originates from the inferior alveolar nerve, there is dilatation of the mandibular canal.

Diagnosis

- **Clinical and radiological diagnosis**—it is difficult to make clinical and radiological diagnosis.
- **Laboratory diagnosis**—biopsy shows plumps of spindle shaped cells arranged in streams and cords with random nuclei. It exhibits variation in degree of malignancy, i.e. tumors that are relatively acellular, show little cellular pleomorphism. Tumors that are highly cellular shows pleomorphism and bizarre mitotic activity.

Management

- **Radical surgical resection**—this is the treatment of choice in neurofibrosarcoma.
- **Adjuvant radiotherapy and chemotherapy**—this can be given in combination with surgical resection.

Muscle Tumors

Leiomyosarcoma

It is a malignant tumor of smooth muscle origin.

Clinical Features

- **Age and sex**—it can occur at any age with no sex predilection.
- **Site**—the most common site is uterine wall and gastrointestinal tract. It is very rare in oral cavity and if present, it is seen commonly on cheeks and floor of mouth.
- **Symptoms**—the lesion appears as painful swelling.
- **Appearance**—the mass is nonspecific in appearance. Secondary ulceration of mucosal surface can be present.
- **Metastasis**—they tend to metastasize through hematogenous route.

Diagnosis

- **Clinical and radiological diagnosis**—nonspecific appearance.
- **Laboratory diagnosis**—there is presence of mitosis, nuclear pleomorphism, hyperchromatism and bizarre cell forms. Proliferating smooth muscles cells are arranged as interlacing bands or cords.

Management

- **Surgical excision**—radical surgical excision should be carried out.
- **Adjuvant radiotherapy**—it should be given in some cases.

Rhabdomyosarcoma

It is a malignant neoplasm of skeletal muscle origin and it is uncommon in oral cavity.

Types (Histological)

- **Pleomorphic**—it occurs most commonly in extremities and in older individuals.
- **Alveolar**—it is found in head and neck region and in extremities, with early age of occurrence.
- **Embryonal**—it is found in the genitourinary tract and in nasopharynx, with cases reported in oral cavity in the upper and lower labial folds.
- **Botryoid**—is a malignant tumor of vagina, prostate and base of bladder in young children.

Clinical Features

- **Age**—it is most common soft tissue sarcoma in children and adolescents. It is exclusively found in males.
- **Site**—it is a mass occurring in any region of the head and neck where striated muscle or its mesenchymal progenitor cells exist. Intraorally, the tonsils and soft palate are most frequently involved.
- **Symptoms**—depending upon of the size of lesion, there may be divergence of eyes, abnormal phonation, dysphagia, cough, aural discharge or deviation of the jaw.
- **Appearance**—typically, it is a rapidly growing soft tissue mass. It forms polypoid fleshy growth beneath the mucous membrane, with club-like extensions at periphery.
- **Signs**—the overlying skin is usually erythematous or telangiectatic.
- **Spread**—it may spread by either lymphatic or hematogenous routes.
- **Metastasis**—the lesions are occasionally ulcerated and may invade the underlying bone and develop distant metastasis.

Diagnosis

- **Clinical**—rapidly growing soft tissue mass with polypoid fleshy growth.
- **Laboratory diagnosis**—in pleomorphic rhabdomyosarcoma, spindle cells are arranged in haphazard manner. The nuclei are situated often in an expanded end of cells. The 'racquet cell', 'Strap-like' and 'ribbon' cells typically show process of long streaming cytoplasm. In *alveolar rhabdomyosarcoma*, epithelium cells

appear to be 'dropping off' from collagen. In *embryonal rhabdomyosarcoma*, four types of cell, i.e. eosinophilic spindle cells, round eosinophilic cells, broad elongated eosinophilic cells and small, round spindle cells with dark staining nuclei and little cytoplasm are present.

Management

- *Local surgical excision*—surgical excision of the tumor should be carried out.
- *Multiagent chemotherapy*—it consists of vincristine, actinomycin D and cyclophosphamide.
- *Postoperative radiation therapy*—radiation therapy should be given postoperatively.
- *Prognosis*—initially, it has poor prognosis. But nowadays due to advent of multimodal therapy, prognosis is improved dramatically.

Alveolar Soft Part Sarcoma

It is rare neoplasm of uncertain histogenesis. It is thought to be of striated muscle origin.

Clinical Features

- *Age and sex*—it is predominantly a tumor of females, occurring usually in teens or early twenties. Occasional cases also occur in older patients.
- *Site*—head and neck region are common site. The orbit and tongue are most common in head neck location.
- *Appearance*—they are usually slow growing, well circumscribed masses.

Diagnosis

- *Clinical*—not possible to diagnose or suspect clinically.
- *Laboratory diagnosis*—biopsy shows group of large polygonal cells which are arranged around alveolar spaces. Cells have abundant granular eosinophils cytoplasm.

Management

- *Radical surgical excision*—due to the high recurrence rate, radical surgical excision is the treatment of choice.

Tumors of Lymphoid Tissues

Hodgkin's Lymphoma

It is lymphoproliferative disorders arising from lymph nodes and from lymphoid components of various organs. It is neoplastic proliferation of lymphopoietic portion of reticuloendothelial system. It involves the cells of either the

lymphocytic or histiocytic series, in varying degrees of differentiation and occurs in an essentially homogenous population of a single cell type.

It was first described by British pathologist, Thomas Hodgkin in 1832. It is characterized by painless enlargement of lymphoid tissue throughout the body.

The exact cause of Hodgkin's lymphoma is not known. Some author suggested that virus like herpes and oncornavirus can cause Hodgkin's lymphoma.

Types (Histological)

- *Lymphocyte predominant*—abundant lymphocytes, few plasma cells, occasional Reed-Sternberg cell, localized involvement of one side of diaphragm and most favorable prognosis.
- *Mixed cellularity*—lymphocytes, plasma cells, eosinophils, easily identified Reed-Sternberg cell.
- *Nodular sclerosis*—sparse lymphocytes, stromal cells, fibrosis and numerous but bizarre Reed-Sternberg cells. It has poor prognosis.
- *Lymphocyte depletion*—lymphocytes, plasma cells, eosinophils with localized involvement.

Clinical Features

- *Age and sex*—it is characterized by a bimodal age incidence, peak one in young adults and the second in the 5th decade of life with equal distribution between sexes.
- *Onset*—the onset is insidious, usually with enlargement of one group of superficial nodes. The cervical lymph nodes are usually the first to be involved but the disease may start in the mediastinal, axillary, abdominal, pelvic or inguinal lymph nodes.
- *Symptoms*—the involved nodes are painless. Generalized weakness, loss of weight, cough, dyspnea and anorexia are seen. Pain in back and abdomen owing to splenic enlargement, due to pressure of enlarged nodes or involvement of vertebrae.
- *Signs*—the lymph nodes are discrete and rubbery in consistency with overlying skin being freely mobile (**Fig. 16-46**). Splenomegaly is usually seen in later stage. Some patients may manifest pruritis.
- *Pel Ebstein fever*—characteristic features of this disease are *Pel-Ebstein fever*, a cyclic spiking of high fever and generalized severe pruritis of unknown etiology.
- *Effect on surrounding structures*—pressure of enlarged lymph nodes on adjacent structures may cause dyspnea, dysphagia, venous obstruction, jaundice and paraplegia.

Clinical Stages (Ann Arbor Staging)

- *Stage I*—involvement of single lymph node region or extra-lymphatic sites.



Fig. 16-46: Enlargement of lymph nodes seen in Hodgkin's lymphoma (Courtesy Dr Milind Chandurkar).

- *Stage II*—involvement of two or more lymph node regions or an extra-lymphatic site and lymph node region on the same side of diaphragm.
- *Stage III*—involvement of lymph node region on the both sides or without extra-lymphatic involvement or involvement of spleen or both.
- *Stage IV*—diffuse involvement of one or more extra-lymphatic tissues, e.g. liver or bone marrow.
- *Subdivision*—all the above stages are subdivided into A and B categories depending on whether they have systemic symptoms such as weight loss, fever, night sweats. A—absence of systemic signs and B—presence of systemic signs.

Oral Manifestations

- *Incidence*—primary jaw lesions are uncommon.
- *Secondary effect*—secondary effect can be seen in oral cavity in the form of infection due to reduced host immune response.
- *Appearance*—it may appear in the oral cavity as an ulcer or a swelling or as an intra-bony lesion which presents as a hard swelling.

Radiographic Features

- *Site*—it is rarely seen in jaws. The common regions are the posterior maxilla and mandible.
- *Appearance*—malignant lymphoma arising in the oral cavity spreads to bone and cause irregular bone loss to the area of the lesion. There are radiolucent areas separated from each other by normal appearing bone which later become confluent, unless treatment is carried out.
- *Margins*—typically, the radiolucent lesions have diffuse ill defined margins which suggest infiltration of bone.
- *Osteoblastic type*—osteoblastic type is uncommon in jaws, but it is seen in the vertebrae and pelvis. In it, there is frank sclerosis with filling of the marrow spaces by bone. It presents as grayness or whiteness which is abnormal. The margins may be well defined and sharp or irregular and trailing off gradually into the normal bone.

Diagnosis

- *Clinical diagnosis*—discrete enlargement of lymph node which is rubbery in consistency with some systemic signs.
- *Radiological features*—there is foci of radiolucency seen in the jaw.
- *Laboratory diagnosis*—it is characterized by replacement of normal lymph node architecture by an admixture of malignant lymphoid cells and non-neoplastic inflammatory cells. Characteristic *Reed Sternberg cells* are present. Multinucleated giant cells are also present. There is also presence of anemia which is normocytic and normochromic. ESR is raised. There is also raised level of LDH.

Differential Diagnosis

- *Non-Hodgkin's lymphoma*—Hodgkin's lymphoma is differentiated from non-Hodgkin's lymphoma by the presence of cells known as Reed-Sternberg cells which have paired as mirror nuclei and prominent nuclei.

Management

- *Radiotherapy*—irradiation treatment 3500-4000 rads/week over the involved region plus all adjacent sites been given in stage I and II. It is also given after chemotherapy, to sites where there was originally bulk disease.
- *Chemotherapy*—it is given in stage III and IV. Usually combination is given. First combination is MOPP, i.e. mechlorethamine (6 mg/m² IV on day 1 and day 8), oncovin which is also called as vincristine (1.4 mg/m² IV on day 1 and 8), procarbazine (100 mg/m²) orally from day 1 to 14) and prednisone (40 mg/m² orally from day 1 to 14). MOPP combination given in six courses with no drugs is given from day 15 to 28. Second combination is ABVD regimen, i.e. adriamycin (25 mg/m² IV bolus on day 1,8 and 14), bleomycin (10 mg/m² bolus on day 1,14), vinblastine (6 mg/m² IV bolus on day 1,14) and decarbazine (375 mg/m² IV bolus on day 1,14). The cycle should be repeated on 20th day.
- *Combination*—a combination of radiotherapy and chemotherapy may increase the overall response and long term survival but, it is associated with delayed complications like leukemia, gonadal atrophy and avascular necrosis of bone.
- *Splenectomy*—splenectomy is advocated in many patients, except with stage IV disease.

Non-Hodgkin's Lymphoma

It is also called as '*lymphosarcoma*'. In this group, there is neoplastic proliferation of lymphoid cells, usually affecting the B-lymphocytes. Unlike Hodgkin's lymphoma, the

disease is frequently widespread at the time of diagnosis, often involving not only the lymph nodes but also bone marrow, spleen and other tissue. Early involvement of bone marrow is typical of this lymphoma.

Types (Histological)

- *Nodular*—neoplastic cells tend to aggregate in such a way that large clusters of cells are seen.
- *Diffuse*—there is monotonous distribution of cells with no evidence of nodularity or germinal center pattern.

Etiology

- *Viral*—the etiology is unclear but herpes virus and Epstein barr etiology has been suggested.
- *Immunological*—there may be induced immunologic effect permitting a malignant clone to proliferate.

Clinical Features

- *Age and sex*—it affects persons of all ages from infants to the elderly. But is commonest in middle age group. Males are affected more commonly than the females.
- *Onset*—the onset of symptoms may be insidious. Painless lymph node enlargement of abdominal and mediastinal region are the most common finding. Very often the first group of lymph nodes affected may be cervical, axillary or inguinal.
- *Symptoms*—the patient complains of tiredness, loss of weight, fever and sweating. Pain is the main symptom of bone involvement which may present as a pathological fracture. Patient may complain of abdominal pain, nausea, vomiting, diarrhea or intestinal obstruction which may occur due to involvement of gastrointestinal tract. Pressure effect of lymphoma may cause dysphagia, breathlessness, vomiting, intestinal obstruction or ascites and paraplegia.
- *Signs*—if liver and spleen are involved, hepatosplenomegaly is present. The growth is fleshy and is prone to ulceration.

Oral Manifestations

- *Site*—occurrence of malignant lymphoma in oral cavity is rare; when present it is more often found to arise from the tonsils. Other sites which can be involved are palate, buccal mucosa and gingiva.
- *Appearance*—palatal lesions have been described as slow growing, painless, bluish soft tissues mass which may be confused with minor salivary gland tumors.
- *Symptoms*—paresthesia of mental nerve has been reported. Sometimes there is pain and neuralgia in the region of 2nd and 3rd division of 5th cranial nerve.

- *Sign*—in rare cases necrotic proliferation of palate may also be seen. The swelling may ulcerate and discolor in some cases.

Radiographic Features

- *Appearance*—as the disease progresses small radiolucent foci scattered throughout the area may be seen. Subsequent radiographs of the expanding lesion will show that these small foci have coalesced to form large multilocular moth eaten radiolucency with poorly defined margins.
- *Margins*—lesion blends imperceptibly with adjacent normal bone, in most of the cases.
- *Effect on surrounding structures*—lesions may cause marked expansion of bone. Erosion and perforation of cortex may occur.
- *Maxillary sinus involvement*—if the lesion involves maxillary sinus, possible opacification with breached cortical walls and associated paracentral or intracentral mass.
- *Teeth*—cortices of unerupted tooth buds and lamina dura of adjacent teeth are lost. Teeth may be resorbed.

Diagnosis

- *Clinical diagnosis*—bluish color mass of the palate with multiple lymph node involvement may suspect the diagnosis of non-Hodgkin's lymphoma.
- *Radiological features*—expansion of bone with radiolucency is present.
- *Laboratory diagnosis*—blood count usually shows hypersplenism or hemolytic anemia. The reduced WBC and RBC counts are seen along with reduced hemoglobin levels and reticulocytosis. In some cases, there may be slight increase in lymphocytes and thrombocytopenia. Moderate degree of anemia will also present when there is considerable bone marrow involvement. Some very aggressive high grade non-Hodgkin's lymphomas are associated with very high urate levels which can precipitate renal failure.

Differential Diagnosis

- *Multiple myeloma*—Bence Jones proteins are present in urine, borders of lesion are usually well defined.
- *Metastatic carcinoma*—history of primary tumor.
- *Ewing's sarcoma*—it occurs in younger age group.
- *Osteosarcoma*—it can be differentiated by clinical features.

Management

- *Chemotherapy*—when diagnosed in late stages, chemotherapy is the treatment of choice. In most cases single agent chemotherapy (chlorambucil) is usually

given. Combination with prednisolone is also useful. A combination of cyclophosphamide, doxorubicin, vincristine, bleomycin with prednisolone is commonly used.

- **Radiotherapy**—radiotherapy is used to treat local disease and the patient is given a total dose of 150 rads spread over a period of five weeks.
- **Transplantation**—studies of autologous stem cell transplantation are in progress.

Primary Reticular Cell Sarcoma

It is also called as '*primary lymphoma of bone*'. Most of cases of primary reticular cell sarcoma arise from non-Hodgkin's lymphoma. It may be caused by immunosuppression and viral agents. Primary lymphoma of bone is usually seen in AIDS patient.

Clinical Features

- **Age and sex**—it can occur in any age group but it is common amongst young adults under the age of 40 years. It is common in male as compared to females.
- **Sites**—it most commonly seen in femur, tibia and humerus.
- **Symptoms**—most common complaint is pain. There is also presence of localized swelling of the involved bone. Patient may also complaint of weight loss, fever, and night sweats.
- **Sign**—regional lymphadenopathy is usually present.

Oral Manifestations

- **Site**—it is not common, but it usually occurs more in mandible than maxilla.
- **Symptoms**—pain is common complaint and usually present for years and more. There is also presence of expansile mass.
- **Sign**—the oral mucosa over the involved bone seldom gets ulcerated but at times, appears diffusely inflamed.
- **Teeth**—the teeth usually become exceedingly loose, owing to destruction of bone.
- **Maxillary lesion**—when the lesion involves maxilla, there may be evidence of expansion of the bone as well as symptoms of nasal obstruction due to superior growth of tumor into the floor of nasal cavity.

Radiographic Features

- **Moth eaten appearance**—there is diffuse radiolucency involving the alveolar bone which may present destruction of the supporting bone of the teeth. It may present a moth eaten appearance.
- **Margins**—in the jaw, the tumor usually gives rise to an area of bone destruction with margins suggestive of infiltration.

- **Periosteal reaction**—there may be periosteal reaction. New bone formation may be linear in character or amorphous and radiating spicules of bone are seen which stand more or less perpendicular to the jaw.

Diagnosis

- **Clinical diagnosis**—a primary focus is single bone. Systemic symptoms with local pain with regional lymphadenopathy will give clue to the diagnosis.
- **Radiological diagnosis**—moth eaten appearance with periosteal reaction is present.
- **Laboratory diagnosis**—primary cells of tumor are identical with that of the soft tissue tumors. There may be inflammatory cell infiltration which can lead to confusion in the diagnosis of the lesion. The individual cells are mixed with both mature appearing lymphocytes and large histiocytoid lymphoblast. Many patients have increased level lactate dehydrogenase.

Management

- **Surgical ablation**—surgical ablation is done.
- **Radiation**—X-ray radiation is the treatment of choice. The 5 year survival rate is between 50 to 60%.

Mycosis Fungoides

It is derived from T lymphocytes. It is also called as '*T cell lymphoma*'. This tumor has got propensity to invade the epidermis of skin (epidermotrophism).

Clinical Features

- **Age and sex distribution**—it usually affects middle aged adult with male predilection.
- **Size**—it varies in size from few millimeters to centimeters in diameter.
- **Site**—it involves lymph nodes, spleen and liver.
- **Eczematous stage**—these are well demarcated scaly erythematous patches. Patient complains of pruritus.
- **Plaque stage**—these appear after eczematous stage in which erythematous patches become elevated red lesion.
- **Tumor stage**—plaque grows and become papules and nodules. In this stage visceral involvement is seen.
- **Sézary syndrome**—it is dermatopathic T cell leukemia. Patient is suffering from exfoliative erythroderma, lymphadenopathy, and hepatomegaly and splenomegaly.

Oral Manifestation

- **Site**—tongue, palate, buccal mucosa, lip, gingiva and tonsils.
- **Appearance**—oral lesions can be the first manifestation and sometimes, appear after the skin lesions have been treated and remitted. The lesions appear as indurated areas, nodules or erythematous ulceration (**Fig. 16-47**).



Fig. 16-47: Indurated area in the maxillary tuberosity region which later on histologically diagnosed as mycosis fungoides.

Diagnosis

- *Clinical diagnosis*—eczematous or plaque lesion on the skin.
- *Laboratory diagnosis*—biopsy shows dense infiltration of lymphoid cells with irregularly shaped nuclei. In some cases, nuclei are so convoluted that they are described as cribriform.

Management

- *Early stage*—topical nitrogen mustard, electron beam therapy and photochemotherapy (PUVA) are used in early stage.
- *Extracorporeal photopheresis*—extracorporeal photopheresis is used in this stage. This involves ingestion of photoactive drug 8-methoxypsoralen followed by removal of patient's blood and separating red and white blood cells. The white blood cells are irradiated outside the body with ultraviolet radiation and then infused back to patient. This will generate immunological response to patient abnormal lymphocytes.

Burkitt's Lymphoma

It was described by Dennis Burkitt in 1950. It is also called as 'African jaw lymphoma'. It is a lymphoreticular cell malignancy. In the African form jaw involvement is 75% and in cases of the American form, abdomen involvement is more common. It is a B-cell neoplasm. Difference between African form and American form is described in **Table 16-2**.

Etiology

- *Epstein-Barr virus (EBV)* which also causes nasopharyngeal carcinoma and infectious mononucleosis is considered to be the etiological factor. There are higher EBV antibody levels in patients of Burkitt's lymphoma.

Clinical Features

- *Age and sex*—peak incidence is in children between 6 to 9 years. Males are affected more commonly than the females, with a ratio of 2:1.
- *Site distribution*—more are found in maxilla than in mandible, where it may spread rapidly to the floor of the orbit. Almost always occurs in molar area. In the African form, more than one quadrant is involved while in the American form, only one quadrant is involved.
- *Onset and progress*—the most important hallmark of this tumor is the fast growth with a tumor doubling time of less than 24 hours.
- *Symptoms*—the most common presenting features are swelling of the jaws, abdomen and paraplegia. It is painless.
- *Sign*—peripheral lymphadenopathy is common.
- *Prognosis*—it is rapidly fatal in the absence of treatment, with death occurring within 6 months.

Oral Manifestations

- *Onset and extent*—it begins generally as a rapidly growing tumor mass of the jaws, destroying the bone

Table 16-2: Difference between endemic (African) and non-endemic (non-African) form

Features	Endemic form	Non-endemic form
Age	Young children	Adolescents and young adults
Nodal tissue	Secondarily involve the extra-nodal tissue	Lymph nodes, lymphoid tissue and bone marrow
Sex	No sex predilection	American male predilection
Jaw	Rapidly growing tumor mass of jaws, destroy bone and cause loosening of teeth	Jaw uncommon
Visceral involvement	Visceral organ involvement is less frequent	Visceral organ involvement is common
Nerve involvement	Maxillary and mandibular nerve not involved	Not common
Antibody titre	Antibody titre is negative	Antibody titre is positive

with extension to involve maxillary, ethmoid and sphenoid sinus as well as orbit.

- **Symptoms**—loosening or mobility of permanent teeth. There is gross distortion of the face due to swelling. Paresthesia and anesthesia of inferior alveolar canal or other sensory facial nerves are common.
- **Signs**—gingiva and mucosa adjacent to the affected teeth become swollen, ulcerated and necrotic. As the tumor mass increases, the teeth are pushed out of their sockets. Swelling of the jaw occurs and it may cause facial asymmetry. They are capable of blocking nasal passages, displacing orbital contents and eroding through skin. There is derangement of arch and occlusion. There may be large quantity of mass protruding into the mouth, on the surface of which may be seen rootless, developing permanent teeth.
- **Spread**—once the tumor perforate the bone, it is initially confined by the periosteum, but subsequently it spreads to the soft tissues of the oral cavity and face where rapid tumor growth soon obliterates the entire face and skin becomes tense and shiny.

Radiographic Features

- **Moth eaten appearance**—small radiolucent foci scattered throughout the affected area. These small foci coalesce and form a multilocular moth eaten appearance. Focal areas of radiolucency are darker and sharper than the shadow of the marrow space lined by normal bony trabeculae.
- **Sunray appearance**—if periosteum is elevated, it will produce sunray appearance.
- **Margins**—margins are ill defined and non-corticated.
- **Shape**—they expand rapidly and are ballooned shaped.
- **Teeth**—Lesions are osteolytic with loss of lamina dura about the erupted teeth and crypts of developing teeth are enlarged. Erupted teeth in the area are grossly displaced, as are the developing tooth crypts.
- **Effect on surrounding structures**—they expand very rapidly and breach its outer cortical limits, causing gross balloon-like expansion with thinning of adjacent structures and production of soft tissue mass adjacent to the osseous lesion. Inferior border of mandible is thinned and later destroyed. Erosion and perforation of the cortex is seen. In some cases, orbit is involved and there is displacement of orbital content.
- **Maxillary sinus**—in the maxilla, there is blurring of shadow of antrum.

Diagnosis

- **Clinical diagnosis**—swelling of the jaw and abdomen with peripheral lymphadenopathy can give clue to the diagnosis.

- **Radiological diagnosis**—moth eaten appearance is seen with loss of lamina dura around the teeth.
- **Laboratory diagnosis**—monotonous sea of undifferentiated monomorphic lymphoreticular cells, usually showing abundant mitotic activity. There is also hyperchromatosis and loss of cohesiveness. Characteristic 'starry sky' appearance is seen.

Differential Diagnosis

- **Non-Hodgkin's lymphoma**—it occurs in older age groups.
- **Cherubism**—it does not breach the bony borders.
- **Osteosarcoma**—it is distinguishable from clinical features. On radiograph, osteoblastic activity is visible.

Management

- **Cytotoxic drugs**—cytotoxic drugs like cyclophosphamide 40 mg/kg in single IV administration and repeated about 2 weeks later. Vincristine and methotrexate have been successful in some cases.
- **Multiagent chemotherapy**—combination of drugs such as cyclophosphamide, vincristine and methotrexate give better results than any single drug. Majority of patients show dramatic response to the therapy. The swelling regresses and the displaced teeth return to their normal position within 1 to 2 weeks.

Leukemia

It is also called as 'leucosis'. It is defined as a neoplastic proliferation of WBC in bone marrow, usually in circulating blood and sometimes in other organs such as liver, spleen and lymph nodes. Presence of leukemic cells in bone marrow results in impairment of normal hemopoiesis with resultant anemia, granulocytopenia and thrombocytopenia. Leukemia is a progressive and fatal condition causing death from hemorrhage and infection. There is presence of excessive number of abnormal cells in the peripheral blood but leukemia is considered a primary disorder of bone marrow.

Types

- **'Stem' or 'blast' cell leukemia**—when the leukemic cells are too immature to be classified as to cell type, the leukemia is termed as 'stem' or 'blast' cell leukemia.
- **Subleukemia**—when the total WBC is normal and leukemic cells are seen in the peripheral blood is termed as subleukemia.
- **Aleukemia**—when no abnormal leukocytes are found in the peripheral blood (i.e. they can be found only in the bone marrow), the term aleukemia is used.
- **Leukemoid reaction**—when the peripheral blood picture in non-leukemic patient resembles that of leukemia, it is called a leukemoid reaction. In this, absolute neutrophil count remains above 30,000/mm³.

Classification

Acute

- *Acute lymphoblastic leukemia*
 - L_1 —Acute lymphoblastic (principally pediatric)—in it, small cells predominate and nuclei are generally round.
 - L_2 —Acute lymphoblastic (principally adults)—cells are heterogeneous in size and sharp in features, nuclei often show cleft.
 - L_3 (Burkitt's)—there is homogenous population of large cells. Nuclei are round to oval with prominent nucleoli.
- *Acute non-lymphoblastic or myeloid leukemia*
 - M_1 —Myeloblastic (without maturation)—myoblasts predominate with distant nucleoli, few granules are present.
 - M_2 —Myeloblastic (with maturation)—myeloblast and promyelocytes predominate and Auer rods are seen.
 - M_3 —Promyelocytic—hypergranular promyelocytes often with Auer rods are seen.
 - M_4 —Myelomonocytic—myelocytic and monocytic differentiation evident, myeloid elements resemble peripheral monocytosis.
 - M_5 —Monocytic—promonocytes or undifferentiated blast.
 - M_6 —Erythroleukemia—bizarre, multinucleated, megaloblastic erythroblast predominate.
 - M_7 —Megakaryocytic—pleomorphic undifferentiated blast cells with anti-platelet antibodies, myelofibrosis are present.

Chronic

- *Chronic lymphatic leukemia* (lymphogenous, lymphocytic) involving lymphocytes series.
- *Chronic myeloid leukemia* (myelogenous, myelocytic) leukemia involving granulocytes series.

Etiology

- *Virus*—Epstein-Barr virus, herpes-like virus and HTLV (human T-cell leukemia virus) have been considered to be the etiological agents responsible for leukemia.
- *Radiation and atomic energy*—if given over the dose of 100 rads, it is known to significantly increase the risk of leukemia. Leukemia among radiologists and Japanese exposed to the atomic blast are more, as compared to other population. It is also common in patients receiving X-radiation for rheumatoid spondylitis.
- *Chemical agents*—chronic exposure to aniline dyes; benzene and phenylbutazone have been recognized to be associated with leukemia. Usually in these patients pancytopenia due to marrow hyperplasia occurs prior to leukemia.

- *Anticancer drugs*—patient treated with anticancer drugs like melphalan and chlorambucil have an increased risk of developing leukemia, usually of acute myelocytic variety.
- *Genetic and chromosomal factors*—Philadelphia chromosome is found in about 15% of cases of acute lymphocytic leukemia. It suggests that if one set of identical twins develop leukemia before the age of 6 years, the risk of disease in other twins is 20%.
- *Immunological deficiency syndrome*—the persons suffering with Wiskott-Aldrich syndrome, Down syndrome, Bloom syndrome, Schwabach syndrome, and Klinefelter syndrome can also develop leukemia.

Acute Leukemia

Acute leukemia is a disorder in which there is a failure of maturation of leukocytes. As a result, there is an accumulation of immature cells within the bone marrow and later in the blood. It is the most common type of leukemia, except in children (in whom acute lymphoblastic leukemia is more common).

Etiopathogenesis

- *Leukemic blast*—there is a block in differentiation of leukemic and stem cells. Thus, accumulation of leukemic blast in acute leukemia results primarily from failure of maturation into functional stage.
- *Suppression of stem cells*—as leukemic blast accumulates in the marrow, they suppress the normal hematopoietic stem cells. The mechanism is not fully understood. Suppression part is related to physical replacement of normal precursor cells by expanded clones of leukemic cells.
- *Clinical manifestation*—clinical manifestations result from paucity of normal red cells, white cells and platelets.

Clinical Features

- *Age and sex*—it is more common in children and young adults between the age of 15 and 39 years. Males are affected more commonly than females with a ratio of 3:2. Acute lymphoblastic leukemia is seen in children.
- *Onset*—there is abrupt stormy onset with pyrexia and enlargement of spleen.
- *Symptoms*—symptoms usually result from bone marrow suppression and infiltration of other organs and tissues by leukemic cells. Weakness, fever, headache, generalized swelling of lymph nodes, petechiae or hemorrhage in skin and mucus membrane are seen. There is also bone pain and tenderness, resulting from marrow expansion, with infiltration of subperiosteum. Central nervous manifestations such as headache, vomiting,

nerve palsies resulting from meningeal spread which are more common in children than in adults; and more common in ALL than AML.

- **Signs**—the clinical features are due to anemia and thrombocytopenia viz pallor, dyspnea, fatigue, petechiae, ecchymosis, epistaxis and melena. Hepatosplenomegaly is present in later stages. There is an increased susceptibility to infection. Cervical lymphadenopathy, secondary to pharyngeal sepsis is seen. Intracranial and subarachnoid hemorrhage may result from thrombocytopenia and leukostasis (intravascular clumping of leukemic blasts in the small blood vessels of brain). There is recurrent infection of lungs, urinary tract skin, mouth, rectum and upper respiratory tract, which may result in fever.
- **Chloroma**—localized tumors consisting of leukemic cells are called chloromas, surface of which turn green when exposed to light because of the presence of myeloperoxidase.

Oral Manifestations

- **Lymph nodes**—the submental, cervical and pre- and post-auricular lymph nodes may be enlarged and tender.
- **Symptoms**—paresthesia of lower lip and chin may be present. There may be toothache due to leukemic cell infiltration of dental pulp.
- **Signs**—the oral mucous membrane shows pallor, ulceration with necrosis (**Fig. 16-48**), petechiae, ecchymosis and bleeding tendency. There may be massive necrosis of lingual mucosa with sloughing.
- **Gingiva**—gingiva shows hypertrophy (**Fig. 16-49**) and cyanotic discoloration. The hypertrophy may be due to leukemic cell infiltration within gingiva or due to local irritants. The gingiva appears boggy, edematous and deep red bleed easily due to ulceration of sulcus epithelium and necrosis of underlying tissue.



Fig. 16-48: Ulceration seen in palate in posterior area in leukemia patient (Courtesy Dr Bhaskar Patle).

- **Teeth**—mobility of permanent teeth may be present.
- **Lip**—in some cases crusting of lip can be seen (**Fig. 16-50**).
- **Infection**—oral infections (candidial, viral and bacterial) are serious and potentially fatal complications in leukemic patients.



Fig. 16-49: Hypertrophy seen in gingiva (Courtesy Dr Milind Chandurkar).



Fig. 16-50: Crusting of lip seen in leukemia.

Diagnosis

- **Clinical diagnosis**—gingival hypertrophy with symptoms of anemia and thrombocytopenia may suspect the leukemia.
- **Laboratory diagnosis**—the total WBC count may vary from a very low count less than 1×10^6 per cu mm to as high as 5000×10^6 per cu mm or more. The peripheral smear shows significant number of immature granulocytes or lymphatic precursors or even stem cells. Bone marrow is hypercellular with replacement of normal marrow elements by leukemic blast cells in varying degree. There may be an associated normochromic anemia, thrombocytopenia and decrease in normal functioning neutrophils count.

Table 16-3: Summary of drugs commonly used in treatment of leukemia

Phase	Lymphoblastic	Myeloid
Induction	Vincristine (IV) Prednisone (oral) A-asparaginase (IV) Daunorubicin (IV) Methotrexate (intrathecal)	Daunorubicin (IV) Cytosine arabinoside (IV) Etoposide (IV) Thioguanine (oral)
Consolidation	Daunorubicin (IV) Cytosine arabinoside (IV) Etoposide (IV)	Cytosine arabinoside (IV) Amsacrine (IV) Mitozantrone (IV)
Maintenance	Prednisone (oral) Vincristine (IV) Mercaptopurine (oral) Methotrexate (oral)	

Management (Table 16-3)

- *Phase of induction*—it is treated using combination of vincristine (1.4 mg/m² every week of 1 month), L-asparaginase (600 units/m² biweekly for 1 month) and prednisone (40 mg/m² orally daily for 1 month). As the leukemic cells regress, regrowth of normal cells occurs and the patient goes rapidly into remission. Acute non-lymphoblastic leukemia is treated with daunorubicin, cytarabine and 6-thioguanine. All these drugs are very toxic to the normal as well as leukemic cells and therefore, treatment is given in pulses to reduce toxicity.
- *Phase of consolidation*—in it, drugs used are daunorubicin, mercaptopurine, cytarabine and methotrexate with intrathecal therapy using the last two drugs, together with irradiation of the cranium to eradicate the disease from central nervous system. Radiation reduces the risk of relapse in central nervous system when given with methotrexate.
- *Phase of maintenance*—in this phase, patient receives a repeating cycle of above drugs until two or three years have been completed. This therapy requires lower dose of chemotherapeutic drugs.
- *Transplantation*—allogenic bone marrow transplantation from HLA identical twin is done. Ablative therapy has been utilized to eradicate the patient's residual leukemic cells and normal cells with intensive irradiation and chemotherapy. After this, reconstruction of the hemopoietic tissue is done with precursor cells from the donor.
- *Allopurinol*—catabolic products of leukemic cells produce uric acid and cause hyperuricemia, which is prevented by allopurinol.
- *Supportive therapy*—transfusion of red cells and platelets may be required in cases of severe anemia and thrombocytopenia. Combination of higher antibiotics like aminoglycosides with cephalosporin, allopurinol, before starting anti-leukemic agents are given to prevent hyperuricemia.

- *Topical treatment for gingival bleeding*—to stop gingival bleeding, remove obvious local irritants by scaling and direct pressure is applied by use of absorbable gelatin or collagen sponge topical thrombin.
- *Management of oral ulcers*—it includes topical antibacterial with povidone iodine solution, chlorhexidine rinses or tetracycline rinses.
- *Psychological support*—it is very important as delusion, hallucinations and paranoia are not uncommon during periods of severe bone marrow failure.

Chronic Myeloid Leukemia

Chronic leukemia is characterized by the presence of large leukemic cells and differentiated WBCs in the bone marrow, peripheral blood and other tissues. It has a prolonged clinical course even without therapy.

It is associated with the presence of a distinctive chromosomal abnormality, i.e. Philadelphia chromosome.

Clinical Features

- *Age*—the disease occurs chiefly between the age of 35 and 60 years.
- *Onset*—the disease may be discovered during routine examination, when splenomegaly or an elevated count is noted.
- *Symptoms*—there may be slowly advancing anemia with loss of weight, prominence of abdomen and discomfort in the left upper quadrant due to splenomegaly. Attacks of acute left upper abdominal pain may develop due to infarction of spleen. Anemia causes weakness, fatigue and dyspnea on exertion.
- *Sign*—as the disease progress, thrombocytopenia can cause petechiae, ecchymosis as well as hemorrhage from the skin and mucous membrane. Liver may be enlarged but lymph nodes are normal.

Diagnosis

- *Clinical diagnosis*—not so specific
- *Laboratory diagnosis*—examination of blood shows a normocytic and normochromic anemia. WBC count is considerably increased and may be between 50×10^6 to 500×10^6 cells per cu mm. Peripheral smear shows mature leucocytes (**Fig. 16-51**) but, few immature forms may also be present. The platelet count is often high initially but with treatment, it comes down.

Management

- *Chemotherapy*—the treatment of choice is chemotherapy using the drug busulphan given orally in a dose of 4 mg daily or in large doses of 50-100 mg spaced 2 to 3 weeks

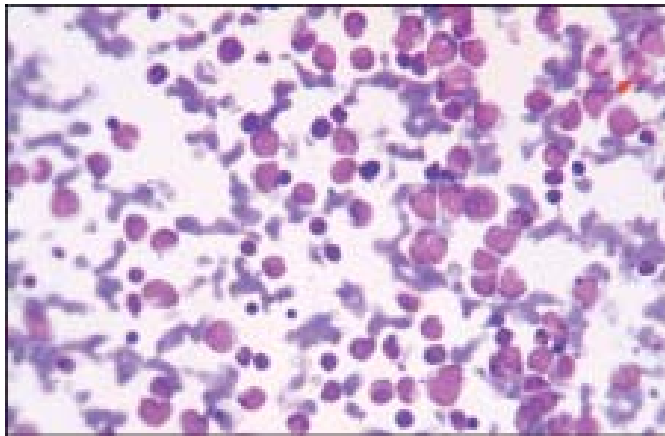


Fig. 16-51: Chronic myeloid leukemia showing mature leukocytes.

apart. The treatment is continued for 12 to 18 weeks and should be discontinued when WBC count is between 10×10^6 and 20×10^6 /cu-mm, otherwise busulphan may cause aplasia of the bone marrow.

- *A combination chemotherapy* using a small dose of busulphan 2 mg daily along with mercaptopurine 50 mg daily or thioguanine 80 mg daily.
- *Other therapy*—radiotherapy and splenectomy are other treatment of choice.

Chronic Lymphatic Leukemia

It is a slowly progressing malignancy involving lymphocytes. It is characterized by the accumulation of long lived, non-functional B lymphocytes.

Clinical Features

- *Age and sex*—it occurs more frequently in males and majority of the patients are over 45 years.
- *Onset*—the onset is very insidious.
- *Symptoms*—tiredness and ill health are common, although some patients are symptom free and the disorder is found incidentally. Bone marrow infiltration causes anemia and thrombocytopenia and results in pallor, weakness, dyspnea and purpura.
- *Signs*—there is a moderate enlargement of lymph nodes which are firm, rubbery and discrete. Liver and spleen are usually enlarged and palpable. There is an increased susceptibility to infection as the leukemic B cells are non-functional. Leukemic infiltration results in skin nodules, intestinal malabsorption, pulmonary obstruction or compression of the central or peripheral nervous system.
- *Lymph nodes*—the most common groups of lymph nodes involved are cervical, axillary and inguinal group.

Clinical Staging

- *Stage A*—Lymphocytosis is less than three areas of lymphoid enlargement, no anemia or thrombocytopenia.

- *Stage B*—more than three areas of lymphoid enlargement, no anemia or thrombocytopenia.
- *Stage C*—anemia or thrombocytopenia, regardless of number of area of lymphoid enlargement.

Oral Manifestations

- *Gingiva*—the most common oral finding is hypertrophy of gingiva. There may be ulceration with necrosis and gangrenous degeneration; a dark brown exudate and foul fetor oris are present.
- *Tongue*—the tongue is frequently swollen and dark.
- *Lymph nodes*—there is regional lymphadenopathy.
- *Teeth*—rapid loosening of teeth due to necrosis of periodontal ligament has been reported. Destruction of alveolar bone also occurs in some cases.

Diagnosis

- *Clinical diagnosis*—signs of anemia and thrombocytopenia with gingival hypertrophy and insidious onset may give clue to the diagnosis.
- *Laboratory diagnosis*—peripheral blood smear shows mild anemia and a large number of small lymphocytes. Lymphoblasts are rare but increase in the terminal stages of disease. WBC count may increase up to 1000×10^6 per cu mm.

Management

- *Supportive treatment*—general measure to maintain good health; adequate rest, good food and exercise should be advised.
- *Chemotherapy*—chlorambucil 6-10 mg/day for 14 days with break of 14 days and cyclophosphamide 2-3 mg/kg IV for 6 days.
- *Combination therapy*—cyclophosphamide doxorubicin, vincristine and prednisone have been recommended.
- *Radiotherapy*—it is useful for large granular masses, if they cause symptoms. Radiotherapy with very small doses, of only 150 rads over a period of five weeks, is very effective and may induce satisfactory remission.
- *Steroids*—if the bone marrow is severely involved, initial treatment with prednisone 40 mg daily and 25-50 mg daily later should be given.

Radiographic Features of Leukemia

- *Site*—it affects the entire body as it is malignancy of bone marrow.
- *Margins*—it is presented as ill defined patchy radiolucent area. This patchy area may coalesce to form larger areas of ill defined radiolucent regions.
- *Chloroma*—foci of leukemic cells may present as a mass which may behave like a localized malignant tumor. It is called as Chloroma.

- *Moth eaten appearance*—destruction of alveolar bone is the most common manifestation of leukemia. Bone loss may be in the form of transverse lines of increased radiolucency or irregular areas and bone loss produced the so called, moth eaten appearance.
- *Sclerosis*—sclerosis of bone may be presented alone or in combination with destructive.
- *Onion peel appearance*—there is formation of bone beneath the periosteum. The new bone takes the form of a thin white line parallel with the shaft of the bone and sometimes separated from it by a thin dark line (onion peel appearance).
- *Skull*—it is rarely involved and it may reveal areas of bone destruction.
- *Teeth*—the developing teeth may be displaced from their normal position.
- *Lamina dura*—there is also loss of lamina dura with loosening of teeth.
- *Jaw bone*—generalized bone loss may be seen.

Differential Diagnosis

- *Metabolic disorders*—some of the metabolic disorders can cause generalized rarefaction of the jaw. But these can be excluded on the basis of blood picture.
- *Lymphoma*—clinical picture and blood testing should be done to rule out leukemia.

Variant of Leukemia

- *Hairy leukemia*—it is a variant of chronic lymphatic leukemia in which there is splenomegaly, severe neutropenia, monocytopenia and the characteristic appearance of hairy cells in blood and bone marrow. These hairy cells appear to be a cross between the lymphocytes and monocytes. It occurs mainly in adults and show male predilection. Manifestations result from infiltration of bone marrow, liver and spleen. Splenomegaly is massive and hepatomegaly is less common. Hairy cell can be identified on the peripheral smear.
- *Prolymphocytic leukemia*—it is another variant of chronic lymphatic leukemia in which there is massive splenomegaly with little lymphadenopathy and a very high WBC count. The characteristic cell is a large lymphocyte with prominent nucleus.
- *Aleukemic leukemia*—it is the sub-leukemic form of leukemia in which the WBC count of the peripheral blood is normal or even subnormal and abnormal or immature leucocytes may be present.

Dental Considerations

- *Topical treatment*—to stop gingival bleeding, removal of local irritants, direct pressure and use of absorbable

gelatin, collagen sponge, topical thrombin and placement of microfibrillar collagen are carried out.

- *Platelet transfusion*—if these local measures are not successful, platelet transfusion is given.
- *Predental treatment consideration*—platelet transfusion and intravenous combination of antibiotics are required before any dental treatment.

Myeloma

Multiple Myeloma

It is also called as '*myelomatosis*'. It is a malignant neoplasm of plasma cells of the bone marrow with widespread involvement of the skeletal system, including the skull and jaws. It is thought to be multicentric in origin.

Origin

- *Proliferation of single clone*—normal plasma cells are derived from B-cells and produce immunoglobulin, which contain heavy and light chain. In myeloma, plasma cells produce immunoglobulin of single heavy and light chain, a monoclonal protein commonly referred as para-protein. There is proliferation of a single clone of abnormal plasma cells in the bone marrow.
- *Bence Jones proteinuria*—in some cases, only light chains are produced and these appear in urine as Bence Jones proteinuria.

Clinical Features

- *Age and sex*—the most common age group affected is between 40 and 70 years with male to female ratio 4:1.
- *Site*—usually the skull, clavicle, vertebrae, ribs, pelvis, femur and jaws are involved.
- *Symptoms*—skeletal pain associated with motion or pressure over the tumor masses, is an early symptom. Pain in the involved bone may be aggregated by an exercise and relieved by rest. Patient may complain of vomiting due to increase in the serum calcium level.
- *Pathological fracture*—spontaneous pathological fracture can also occur.
- *Bleeding tendency*—patient may have bleeding tendency and bruising of skin due to anemia and thrombocytopenia. The cause of bleeding is that the abnormal globulins bind with coagulation factors which also increase the viscosity of blood.
- *Sign*—swelling over the areas of bone involvement may be detectable. There is an increased susceptibility to infection due to abnormal immunoglobulin production by the plasma cells.
- *Hypercalcemia*—mobilization of calcium from the skeleton may cause hypercalcemia resulting in

nephrocalcinosis, lethargy, drowsiness and eventually coma, if untreated.

- **Complication**—the common cause of death is renal failure, caused by accumulation of abnormal proteins in the renal tissue.

Oral Manifestations

- **Site**—mandible is more commonly involved than the maxilla and particularly angle of the mandible, because of its greater content of marrow. Lesions have also been reported in temporomandibular joints.
- **Symptoms**—the patient may experience pain, swelling and numbness of the jaw. Epulis formation or unexplained mobility of teeth is also detectable.
- **Signs**—intraoral swelling tends to be ulcerated, rounded and bluish red similar to a peripheral giant cell lesion. Sometimes, swelling may erode buccal plate and produce rubbery expansion of jaw. Chronic trauma produces an inflamed and ulcerative necrotic surface. On palpation, swelling is tender and eggshell cracking may be elicited.
- **Signs of bone marrow involvement**—secondary signs of bone marrow involvement such as pallor of oral tissue, intraoral hemorrhage and susceptibility to infection may also be seen.
- **Hemorrhage**—excessive hemorrhage may be caused by thrombocytopenia, secondary to increased proliferation of the plasma cells in marrow.
- **Oral amyloidosis**—it is a complication of this disease. The tongue may be enlarged and studded with small garnet-colored enlargements, including nodes on lip and cheeks. Tongue enlargement may cause impairment of speech, mastication and deglutition. Amyloid can also deposit in the gingivae, where it can cause soreness and inability to wear dentures.
- **Oral hairy leukoplakia and candidiasis**—this can be present in rare cases of multiple myeloma as myeloma is also immunosuppressive disease.

Radiographic Features

- **Sites**—they are usually bilateral and may become confluent forming a larger area of destruction. It is seen in mandibular posterior region and ramus.
- **Punched out lesion**—in the radiograph (**Fig. 16-52**), it appears as a small rounded and discrete radiolucency having punched out appearance. There may be numerous areas of bone destruction within the region of generalized radiolucency. Diffuse destructive lesions of bone may occur.
- **Shape**—some lesions have oval and cystic shape.
- **Margins**—it has well defined margins and lack the circumferential osteosclerotic activity. In some cases,

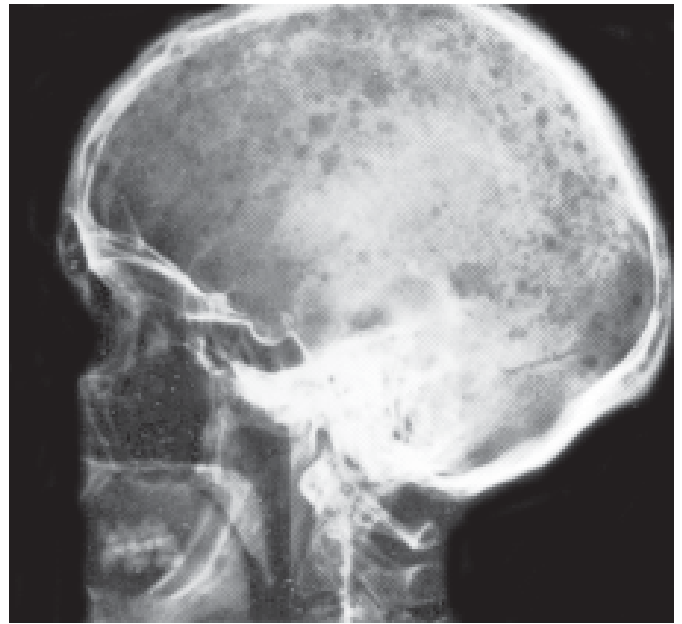


Fig. 16-52: Punched out lesion seen in skull of patient of multiple myeloma (Courtesy Dr Ganju).

borders of the lesion may display a thin sclerotic rim and even areas of opacification.

- **Size**—the lesions may vary in size from few millimeters to centimeters, or more in diameter.
- **Soft tissue lesions**—soft tissue lesions have been reported in jaws and nasopharynx, which appear on radiograph as smooth bordered soft tissue masses, possibly with underlying bone destruction.
- **Teeth**—if good deal of mineral is lost, the teeth may appear to be too opaque and may stand out conspicuously from their osteopenic background.
- **Lamina dura**—lamina dura and follicle of impacted teeth may loose their typical corticated surrounding bone in a manner analogous to that seen in hyperparathyroidism.
- **Cortical border**—mandibular lesion may cause thinning of cortical lower border or endosteal scalloping.

Diagnosis

- **Clinical diagnosis**—skeletal pain with oral amyloidosis with bleeding tendency may give clue to the diagnosis.
- **Radiological features**—punched out lesion in the skull is typical of multiple myeloma.
- **Laboratory diagnosis**—plasma cells are round or ovoid cells with eccentrically placed nuclei exhibiting chromatin clumping in 'cartwheel' or 'checkerboard' pattern. Russell bodies are common finding in multiple myeloma. *Bence Jones proteins*, which coagulate when the urine is heated at 40 degree Celsius, can be demonstrated in the urine of patients suffering from multiple myeloma.

Differential Diagnosis

- *Metastatic carcinoma*—multiple myeloma is, comparatively, more common than metastatic carcinoma. The absence of definitive serum, characteristic history of treatment for an earlier tumor indicates that the disease is metastatic carcinoma.
- *Histiocytosis X*—the incidence of Histiocytosis X is most common in children. The lesions of this disease have ragged and vague margins.
- *Cherubism*—multiple radiolucent lesions of cherubism may be scattered throughout the jaws, commonly seen in the first or second decades of life. Radiolucent lesions may be multilocular or unilocular, elliptical without well circumscribed or hyperostotic borders.
- *Advanced osteomyelitis*—shows the presence of visible cause.
- *Simple bone cyst*—it may be bilateral but they are usually corticated in part and characteristically interdigitate between roots of teeth in much younger population.

Management

- *Chemotherapeutic agents*—general disease is treated with chemotherapeutic agents like melphalan and cyclophosphamide.
- *Management of anemia and hypercalcemia*—patients who present with anemia, hypercalcemia, and evidence of renal damage require urgent management with alkalization of urine with oral bicarbonates, high fluid intake, corticosteroids and possibly mithramycin to reduce calcium level.
- *Blood transfusion*—it may be required if hemoglobin is less than 10 gm/dl.
- *Cell transplantation*—stem cell autotransplant may improve quality of life and prolong survival.
- *Radiotherapy*—this is effective for localized pain not responding to simple analgesics and for pathological fractures.
- *Bi-phosphonate therapy*—it may reduce bone pain and skeletal events.
- *Others drugs*—alpha interferons may prolong the plateau phase. Inorganic fluoride phosphate to reduce bone pain.

Solitary Myeloma

It can appear in any bone and remains for long period of time. It usually involves single bone. The hematological and biochemical changes seen in multiple myeloma are not seen in solitary myeloma. Patient is not anemic and serum protein level is normal. Others symptoms are same as in multiple myeloma.

Plasmacytoma

Plasmacytoma is unifocal, monoclonal and neoplastic proliferation of plasma cell arising within the bone. Extramedullary location without bony involvement may occur in the nasopharynx, nasal cavity, paranasal sinuses and rarely in the oral cavity. It is also called as '*plasma cell myeloma*'. It is least aggressive part of plasma cells neoplasm.

Clinical Features

- *Age and sex*—mean age of occurrence is in age of 51 years and males are affected more commonly than females.
- *Site*—nares, tonsil, palate tongue, gingivae and the floor of mouth. They are also found in pleura, mediastinum, thyroid, ovary, intestine, kidney and skin.
- *Symptoms*—pain and swelling are the most common complaints.
- *Signs*—pathologic fractures are common.
- *Metastasis*—regional metastasis may develop in small number of cases.

Oral Manifestations

- *Site*—it is rare in jaws and can occur in mandible or maxilla.
- *Appearance*—the lesions are described as sessile or polypoid reddish masses in mucous membranes, which become lobulated as they enlarge but exhibits the tendency to ulcerate.
- *Symptoms*—pain is not prominent symptom unless bone is invaded.
- *Signs*—there may be bleeding and ulceration of oral mucosa.

Radiographic Features

- *Forms*—it mainly occurs in two forms, one is purely destructive lesion suggestive of metastatic carcinoma and the other type is an expansible lesion, suggestive of a giant cell tumor.
- *Destructive lesion*—there is no new bone at the margins or under the periosteum. The margins are clearly defined or show evidence of infiltration. In the flat bone, solitary myeloma extends along the bone and produce a saucer-shaped lesion, with undulating margins.
- *Expansible lesion*—an area of bone destruction is well demarcated from the surrounding bone and sometimes is corticated. If the bone is expanded, subperiosteal new bone may form a complete bony covering of the tumor. In the substance of the lesion, trabeculae vary considerably in number and thickness. Some are thin and delicate, while others are coarse.

Diagnosis

- *Clinical diagnosis*—it is more or less mimicking multiple myelomas.
- *Radiological features*—destructive lesions are seen in jaws.
- *Laboratory diagnosis*—it contains densely packed plasma cells and is indistinguishable from the bony lesion in multiple myeloma. There may be considerable nuclear and cellular pleomorphism. Russell's bodies can also be found. It is similar to multiple myeloma. Bence Jones proteins are found in urine. There is also hyperglobulinemia and anemia.

Management

- *Radiotherapy*—radiotherapy at dose of 4000 cGy is given.
- *Surgical excision*—in some cases, conservative surgical excision should be carried out.

Malignant Tumors of Salivary Gland

It is described in Chapter 26: Salivary Gland Disorders.

Treatment Modalities of Oral Cancer

Oral cancer is treated by surgery, radiotherapy and chemotherapy or by combination of these. The other developing techniques in the treatment of oral cancer are laser therapy, hormonal therapy, hyperthermia, etc.

Following histological confirmation of oral cancer, each patient should undergo a thorough assessment. First, the extent of the malignant disease should be accurately delineated and also, the fitness of the patient to undergo treatment is determined. Many patients afflicted by this disease are elderly; such patients likely to be suffer from other chronic illness and also are at risk of having malignant diseases of respiratory or digestive tract.

The oral lesion is carefully examined and the details are recorded. Photography is of great value in assessing the response of the lesion to treatment. The degree of involvement of adjacent structures by tumor can be assessed by palpation.

Radiotherapy

Advantage of Radiotherapy

- *Avoiding surgical procedure*—due to radiotherapy, major surgical procedure is avoided.
- *No cosmetic and functional defect*—as surgery is avoided, there is no cosmetic or functional defect.
- *Management of early metastasis*—early metastatic spread to cervical lymph nodes can be treated.

- *Management of multiple lesion*—multiple malignant lesions at different anatomic sites can be treated at the same time.

Disadvantages

- *Osteoradionecrosis*—there is risk of post-irradiation osteoradionecrosis.
- *Effect on normal tissue*—there is harmful effect on soft tissue and salivary glands.
- *Retreatment not possible*—in case of failure or recurrence, re-treatment with radiation is not very successful.

Indications

- *Vascularized lesion*—it is indicated in early well vascularized lesion.
- *Cervical lymph nodes*—elective irradiation of the cervical lymph nodes.
- *Post-surgical*—to destroy malignant cells peripheral to a large lesion after surgical procedure.
- *Palliative treatment*—palliative radiotherapy for a large lesion, which is not surgical irradiation.

Types of Beams

Electron Beam

X-ray is produced in an X-ray machine first by accelerating electron and then making them strike a target. This causes the electron to loose energy in the form of X-ray photon which is directed towards the patient, for either diagnostic or therapeutic purpose.

The energy of the photon is proportional to the energy of the electron from which they were produced which in turn is dependent on the speed with which electrons were travelling before they struck the target.

Proton Beam

Protons are much heavier than electrons and have a positive charge. They have a finite range like electrons but at the end of their range, Bragg's peak occurs where they deliver the maximum dose.

Protons have virtually no side scatter which adds to the therapeutic ratio. By appropriately adjusting the proton beam, Bragg's peak can be positioned with the tumor almost anywhere in the body, sparing the normal tissues; both proximal and distal to the tumor.

Neutron Beam

Cancer cells that are hypoxic or that are in certain stages of the cell cycle or that are proficient at repairing damage are relatively resistant to be killed by photon or electron beam irradiation.

It was found in the laboratory that such cells are less resistant to be killed by neutron beam because neutrons produce much denser ionization in tissues, inflicting a relatively greater injury to malignant tissues than the healthy tissues, as compared with photon and electron irradiation.

Radiotherapy Technique, Planning and Equipment

There are three major modalities of radiation therapy—external beam radiation or teletherapy, brachytherapy and radioactive isotopes.

Teletherapy

This is the most common modality of radiation therapy. The X-rays of different energies are used for diagnostic and therapeutic purposes.

Superficial X-ray

These rays have energy between 50 Kilo Voltage (KV) to 140 KV with a limited range of penetration. The tissues deeper than 5 cm receive less than 20% of the dose. Thus, this can treat only superficial skin lesions. The absorption of superficial X-rays is by the photoelectric effect.

The superficial X-rays are used to treat the skin lesions of limited thickness, often with a lead cutout to delineate the field. Only direct field is used with a filter of aluminum to decrease the amount of very low energy X-rays. The quality of X-rays used depends on the maximum energy and filtration used.

Orthovoltage X-ray

These X-rays have energies between 250 KV to 350 KV and these are used as most common treatment modalities. The tissue penetration at 5 cm in depth is about 60%. The maximum absorbed dose from single field is at the skin surface and the deeper tissues are treated by using multiple therapies. Orthovoltage radiations are still used for palliative therapy where poor tissue penetration is of little importance.

Cobalt 60 Unit (Fig. 16-53)

This unit has a cobalt source surrounded by a protective shielding with a shutter system which allows the patient to be treated through an adjustable collimator. The source produced gamma rays of 1.17 MeV to 1.33 MeV which behave exactly like X-rays and have a decay half life of 5.27 years. Hence, the treatment times have to be adjusted monthly. Source, diameter is about 2 cm, which is larger than a linear accelerator target, so the cobalt beam has a large penumbra. The maximum depth at which dose delivered is 5 cm beneath the skin, with 55% of dose

delivered at 10 cm. The disadvantages compared to linear accelerator are decreased tissue penetration and less skin sparing and wider penumbra.



Fig. 16-53: Cobalt 60 unit used to give radiotherapy

Megavoltage

It includes linear accelerators (Fig. 16-54) and betatrons. These machines accelerate electrons across millions of volts to provide a continuous X-ray spectrum, with the maximum energy equal to the accelerating X-ray. These machines normally operate at 4-8 MeV range and spare the skin.

The target in linear accelerator is small, which produces a narrow penumbra compared to that of the cobalt unit. It is more expensive to maintain (than the cobalt unit) and requires staff trained in physics, as it requires daily calibration. It produces high energy beams with good tissue penetration. The energy from these high energy beams is absorbed by Compton Effect. The energy absorption is not dependent on the average weight of the tissues. So, the bones, cartilage and soft tissues receive the same dose and



Fig. 16-54: Linear accelerator used in radiotherapy unit.

thus, reduce the chances of necrosis. Higher the energy of X-ray, greater is the degree of skin sparing and tissue penetration.

Advantages of megavoltage over orthovoltage therapy

- *Less chances of osteoradionecrosis*—orthovoltage irradiation is preferentially absorbed in bones that is why bone stand out from soft tissues on the diagnostic X-ray picture. After irradiation, healthy bone receives a higher dose of radiation than the tumor itself. Therefore chances of osteoradionecrosis of mandible is relatively common with orthovoltage treatment and in megavoltage irradiation dose to bone is no higher than the dose to the tumor and the incidence of osteoradionecrosis is decreased dramatically.
- *Skin sparing*—super voltage photon deposits their maximum dose deeper in the body than the skin while orthovoltage photon deposits maximum dose in the superficial surface of the skin. Higher the energy of super voltage photon, deeper will be the region of maximum dose and this is called as skin sparing and hence the skin reactions are very much less severe with super voltage irradiation.
- *Sharper penumbra*—when the photons enter the body, they interact with atoms which make them scatter. Orthovoltage photons often scatter sideways which means they tend to get out of the line of radiation beam and then end up irradiating adjacent healthy tissues. In megavoltage therapy, there is minimum scattering of photons sideways thus sparing the adjacent tissues. This is important in treating the tumor situated near the eyes and brainstem.

Electrons (4-20 MeV)

Photons have no electrical charge hence they deposit dose in tissue at an exponential rate, which means that tissue beyond the tumor also gets significant irradiation. Electron has a negative charge. A charged particle deposits uniform dose for a certain distance and then virtually none, which means that the tissue behind the tumor gets little irradiation. Thus, the electron beam is better suited than photon beam for treating a tumor located on or just under the skin such as basal cell carcinoma or in metastases to cervical lymph nodes.

High energy photons can deliver a relatively low dose to the healthy tissue proximal to the tumor, while electrons deliver a low dose to the distal tissue. In many situations combining the two can provide the patient with the best dose distribution.

Linear accelerator or betatrons can produce a therapeutic electron beam by allowing the electron to pass through a scattering foil rather than striking a target with X-ray production.

Field Arrangement

Single Field Arrangement

In this, tumor is treated with one field only. These treatments are palliative because of quick and easily reproducible therapy. If the maximum applied dose is to be at the skin surface, wax build up is required. The isodose lines can be corrected by a tissue compensator; the simple compensator is called as a *wedge compensator*, which is a high density metal in the shape of a wedge. Wedge shifts the isodose by decreasing the transmission of X-ray beam with increasing thickness of wedge.

Parallel Opposed Fields

It is the next simple field arrangement where the two parallel fields are opposite to one another. This produces a fairly even distribution throughout the irradiated area.

Wedge Lateral Field

Two right angled fields can be combined to irradiate a discrete tumor volume rather than placing them parallel. The equal distribution is achieved by the use of a field wedge.

Multiple Field

Head and neck tumors are superficial and can be irradiated by two fields, without compromising the depth, while tumors of thorax and abdomen require 3 to 4 fields to achieve an adequate concentrated tumor dose.

Most common form of multiple field treatment is with 4 fields, arranged two parallel and two opposed pairs (4 field brick technique)) which produce good tumor localization with a square volume.

Rotational and Arc Therapy

These are moving field techniques which are modifications of multiple field therapy. In rotational therapy, the radiation source rotates around the tumor isocentrically to provide an even dose throughout the tumor volume with a dose decreasing toward the periphery; this produces excellent tumor localization but all tissues are included in the primary beam. So to spare a selected normal tissue from the primary beam, only part of a rotation (arc therapy) is made.

In arc therapy, wedges are usually required and they may need reversing during treatment, such therapy is time consuming and less frequently used. With the advent of high energy X-ray machines which are able to provide a good dose distribution with 3 to 4 fields, use of arc therapy has declined.

Radiotherapy Planning

The following factors should be considered while planning for radiotherapy:

Patient Positioning and Immobilization

Before planning radiotherapy, patient's position should be chosen which depends on many factors such as treatment machine, general condition of the patient and whether the treatment is radicular or palliative. Radiotherapy can be delivered in supine, prone or even sitting position.

The sitting position is useful in the palliative treatment of bronchial tumors, where a patient is unable to lie flat due to pain. Although the sitting position is well tolerated, better immobilization accuracy and reproducibility are achieved in supine position or prone position. Ideally, the patient should remain in the same position for all fields, with the same support and immobilization device so that the treatment is easily reproduced.

The planning couch must resemble the treatment couch in every way as the patient contour changes on different surfaces. The planning couch must also have the same immobilization as the therapy machine, which normally consists of a range of head rest, arm rest and mouth bites.

Immobilization of the patient of head and neck tumor is essential as high dose must be given accurately to a small volume avoiding the adjacent radiosensitive organs such as brainstem, eye and cervical cord.

A thin transparent plastic shell is used which can be molded to the patient's skin following the surface contour. The field size, entry and exit point, can be redrawn on the shell rather than the patient's skin. The shell usually helps to maintain the patient position during the treatment with the maximum support and comfort.

Simulation and Beam Verification

A simulator is an isocentrally mounted diagnostic X-ray unit that mimics the geometrical arrangement of any therapy machine. A simulator couch is identical to the therapy couch so that the patient maintains the same position.

The patient is placed on the couch in the treatment position and the anterior and posterior field are aligned. This is done either clinically or with the help of fluoroscopic screening facility, using a skeletal reference point. Contrast media and metallic markers can be used to identify the skin and other soft tissues.

The tumor volume is localized in the coronal plane and a verification X-ray is taken. A radiopaque ruler or ring is introduced to calculate the magnification. For single and parallel opposed fields, this is sufficient and the field can

be marked on the patient's skin or shell. Where a tumor volume is required, a lateral film is taken through the treatment plane which determines the anterior and posterior limits in the sagittal plane.

Tumor Volume

After the field position has been verified an outline of the patient is taken through the center of the volume by applying a pliable lead strip to the patient. This outline is then transferred to a paper, with the field in center, the patient in the midline of the couch surface and often a reference point marked.

The tumor volume is calculated from orthogonal X-ray films and put onto the transverse outline through the tumor center. The coronal projections of tumor volume are normally obtained from the anteroposterior films. The lateral check films are used to calculate the sagittal tumor volume. Tumor volume depends on several factors, one of which is the experience and philosophy of the radiotherapist.

Isodose Distribution

A treatment plan is a result of the addition of Isodose curves of the entire field. This will produce an even dose across the tumor volume with less than 5% variation. The dose of surrounding tissues should be as small as possible.

Brachytherapy

In brachytherapy or intracavitary radiation therapy, the radioactive source is close to the patient's body as it is contained in a custom fabricated carrier device and placed directly on the surface of the tumor. Brachytherapy is divided into three types:

Mould Treatment

The radiation source is placed into a plastic mould on the patient's skin or mucous membrane to treat superficial tumors. Most common sources are iridium-192, cobalt-60, caesium-137 and gold-198. The radiation dose from the mould falls off rapidly obeying an inverse square law so that the treatment depth is limited. Deep lesions cannot be treated by this method.

Interstitial Therapy

It involves the insertion of a radioactive source into the tumor.

Intracavitary Therapy

In it, radiation source is placed into the body cavity to irradiate the surrounding tissues. The dose from the source

obeys the inverse square law, so that the dose distribution falls off rapidly.

Radioactive Isotopes

This form of treatment involves the radioactive isotopes that are not confined in a protective container. The useful therapeutic effects can be obtained if the target organ concentrates the isotopes and increases its specific dose. The dose can be made to remain in the body cavitations by chelating them to high molecular weight colloids; the colloids will remain in the body cavity and deliver a high surface dose with little penetration.

Iodine-131 (^{131}I)

It is concentrated in thyroid follicular cells which can be killed by I^{131} hence, it is useful in the treatment of thyrotoxicosis and follicular adenocarcinoma. Physical half life is 8 days but the biological half life is only 3 days. Iodine-131 is given orally and the maximum dose is limited to 200 mCi.

If the patient contains more than 30 mCi of I^{131} , he should be confined in an isolated room and the public contact is allowed only when they got less than 15 mCi of I^{131} , as iodine is excreted in all body fluids. Precaution such as special toilet facilities should be taken.

Phosphorus-32 (^{32}P)

It is taken up by bone so, it can be used in the treatment of polycythemia vera. These isotopes are pure beta emitters and are not excreted readily in body fluids. Radioprotection is much easier and doses above 10 mCi are rarely necessary.

Yttrium-90 (^{90}Y)

The isotopes are pure beta emitter and are available as silicate colloid. It can be injected into the body cavity to palliate malignant effusion. The surface dose is high but falls to 50% at 1 mm depth.

Factors that Influence the Dose Required for Tumor Control

Histology of Malignant Cell

The tumors originating from the cell which are sensitive to radiation are easy to cure while those originating from those cells which are resistant or less sensitive to radiation are difficult to cure, e.g. lymphoma is easier to cure than carcinoma which in turn is easier to cure than osteogenic sarcoma.

Primary Site of the Tumor

If the tumor is located in the tissues which are in close proximity with glottis, brainstem or spinal cord, it becomes difficult to treat such tumors as these tissues get the same exposure to radiation therapy inviting complications. If the tumor is in the region of rich lymphatic drainage, it is difficult to treat and vice versa.

Size of the Tumor

Larger the tumor, higher the dose required to control it. It may be due to ratio between the number of cancer cells (tumor burden) requiring sterilization and the number of ionizing particles (dose) available to destroy them. The treatment depends on the tumor burden, number of hypoxic foci in the tumor and heterogeneity of the tumor. Larger tumors have more hypoxic foci than smaller ones. As the size of the tumor increases, it undergoes more mutation and the tumor becomes increasingly heterogeneous, which also requires to increase the dose for tumor control.

Strategies for Improving the Therapeutic Ratio

The ideal machine for radiotherapy should be that, which deliver a lethal dose to all malignant cells, but without any dose to healthy cells does not yet exist. Therefore, a number of strategies have been developed or are being investigated in an effort to obtain a favorable therapeutic ratio. They are as follows:

Altering the Degree of Radiation Injury

Effort has been made to increase the degree of injury to malignant cells that produced by a given dose of irradiation. The technique for this includes—

Hyperthermia: It selectively injures to two kinds of cells that are most resistant to irradiation, hypoxic cells and cells in the late 'S' phase of the cell cycle. In treating tumors in human beings, eliminating these cold and hot spots has been a major problem. Radiofrequency, microwaves and ultrasound are all being tried for heating human tumor and the technology is rapidly developing.

Hyperbaric oxygen therapy: The most important modifier of the biologic effect of ionizing radiation is molecular oxygen. Oxygen affects the initial chemical product of radiation interaction with biologic material. The important free radicals have shorter half lives and these are very reactive molecules.

It has been shown, from various studies, that oxygen enhancement ratio, i.e. the ratio of dose required for equivalent cell killing, in absence of oxygen, compared with

presence of oxygen ranges from about 2.5 to 3.5 for given survival rate. Three times as much radiation is required under hypoxic conditions than under conditions when oxygen is present.

Neutron therapy: Cancer cells that are hypoxic in certain stages of cell cycle are proficient at repairing damage and relatively resistant to be killed by photons and electron beam irradiation. These cells are resistant to neutron beam because neutrons produce much denser ionization in tissues. So, neutron irradiation inflicts greater injury to malignant tissues than healthy tissues, when compared with photon or electron irradiation.

Radiation protector: Chemicals that protect normal tissue from radiation injury have been sought by radiation oncologist. This compound should have a property of selectively protecting the healthy cells and not the malignant cells. Amongst them, a compound designated WR-2712 was the most promising in this regard.

Optimizing the Distribution of Radiation Dose

During irradiation of a tumor, the neoplastic tissues should receive high dose while the healthy tissues should receive a selectively low dose. To achieve this, following measures are used:

- *Electron beams* are used to treat the deep situated tumors located in skin, cervical lymph nodes metastases.
- *Super-voltage photons* are used to treat the deep situated neoplasms due to their skin sparing effect.
- *Multiple field technique* is used to treat the tumors of deeper tissues. Combination of two or more beams permits more uniform irradiation of the tumor, as compared with a single beam technique.
- *Shrinking field technique*—the malignant cells that constitute the wide peripheral extension of a tumor can be irradiated by a lower dose of irradiation because these cells are oxygenated. The cells constituting the central core of the tumor are more hypoxic and therefore require more doses of irradiation. So, the margins of healthy tissues should be treated to encompass the occult peripheral regional extension along with the known tumor volume. After a certain dose, the tumor mass regresses and the field can shrink to encompass only the central core of the tumor where the dose of radiation is increased. Hence, the surrounding healthy tissues are protected from higher dose of radiation and thus are at reduced risk of complications.
- *Radiolabelled antibodies*—antibodies against a protein specific to the tumor are generated in animals and then labelled with a radionuclide. When these antibodies are injected into a patient, they lodge over the tumor and expose it to irradiation from radionuclides. By this,

extremely high dose of radiations to tumor are possible with virtually no exposure to healthy tissues. But finding of the protein that is specific only to the tumor is difficult.

Optimizing the Timing of Irradiation

Hypofractionation

In it, the same total dose is delivered with a reduced number of fraction, e.g. 6000 rads in 10 fractions of 600 rads each, instead of 30 fraction of 200 rads each. But, this increased dose per fraction will reduce the total dose tolerance, producing a significant increase in the injury to late reacting healthy tissues and the total dose has to be reduced, which compromises the tumor control. So, hypofractionation is not recommended unless low doses to the tumor are acceptable, usually for palliative purpose or unless the patient has such a limited life expectancy that the injury to late reacting tissue is not of concern.

Hyperfractionation

If the same total dose is delivered with an increased number of fractions, it is called as hyper-fractionation. The minimum intervals between fractions that allow reasonable recovery of healthy tissue are 5-6 hours.

Accelerated Fractionation

Most of the tumors have intermitotic intervals of few days like acutely reacting tissue. The dose which increases injury to acutely reacting tissue will increase the injury to malignant cells. When the dose more than 200 rads per day are used in a single fraction, the injury to the late reacting tissues will increase. So, the accelerated radiation must be delivered in multiple conventional fractions per day with 5-6 hours gap between successive irradiations. Thus, in the accelerated fractionation, neither the fraction size is reduced nor increased but, there is an increase in radiations per day, normally with intervals of 5-6 hours.

Combined Strategies

A large number of possible permutations and combinations of hypofractionation, hyperfractionation and accelerated fractionations to treat the head and neck cancers are under investigation.

Reducing the Tumor Burden

Surgery helps to reduce the tumor burden to a level that can be safely irradiated by irradiation. Chemotherapy also helps in reducing the tumor burden in cases where the tumors are not resectable but morbidity of resection is more.

Complications of Radiotherapy

Osteoradionecrosis

Changes in the jaw bone after irradiation results from damage to the bones as well as impaired vascularity. The ready access of a highly varied microbial flora to heavily irradiated tissue is an important factor in the production of osteoradionecrosis. Gross necrotic lesion always begins in the interdental papilla of mandibular molar that was in the center of the field of radiation. The necrosis then extends to cervical gingiva, attached gingiva and the cheek mucosa. Patient may experience a transient prospective or thermal sensitivity of the teeth or perhaps a continuous dull toothache. Spontaneous death of pulp in an erupted teeth has also been reported. Pain in the jaw may be continuous and severe. After varying period of time, radiographic changes may appear. All trauma should be avoided to minimize the possibility of osteoradionecrosis. Deep gingival curettage and extraction are common contraindications in these cases.

Osteoradionecrosis is more easily prevented than treated. Whenever possible, the jaws, teeth and oral mucosal tissue should be shielded before radiation therapy in the area, unhealthy teeth are removed before radiation therapy. As a general rule, teeth which are in direct line of radiation should be rendered healthy enough to require little or no treatment for 5 years after radiation. Surgical intervention is contraindicated, except for the careful removal of sequestra that develops. Antibiotic therapy may reduce secondary infection. Hyperbaric oxygen therapy is also useful in these cases.

Radiation Mucositis (Fig. 16-55)

It is the inflammation of the oral mucosal membrane following radiotherapy. The basal layer of oral mucous membrane contains differentiating intermitotic cells.



Fig. 16-55: Mucositis occur due to radiotherapy.

These cells are sensitive to radiation. It develops in second week of radiotherapy. There are areas of redness and inflammation.

Later on, the mucous membrane breaks down to form pseudomembrane. After some period, mucous membrane becomes atrophic and thin. It is managed with application of topical anesthetizing agents for relief of pain, most commonly used is dyclonine hydrochloride.

Surgery

The surgical treatment of oral cancer as a primary modality is ablative (excisional) in nature. All the clinically detectable tumors must be excised with adequate margins of adjacent normal tissue, to ensure that the residual elements of microscopic disease do not remain within the surgical field. Different types of surgeries may be performed as follows. Surgical treatment has two distinctive phases: the resection of the tumor and the reconstruction of the defect.

Enbloc Resection

In it, removal of the entire tumor along with adequate margins of surrounding normal tissue in continuity, as one intact specimen is carried out, without incision through the involved tissue. Localized tumors without apparent or suspected cervical lymphatic involvement are treated by this method. Skin incisions are necessary to gain sufficient access for larger lesion. Resection should proceed along anatomical planes, whenever possible.

The extent of resection is determined by the margins of carcinoma or potentially involved tissue and is not spared to facilitate reconstruction. The surgeon must clear the site of the tumor at the first attempt. The chance of resecting residual tumor in a second operation is likely to be poor, as the demarcation between normal and pathological tissue becomes obscured by scarring and distortion of the anatomy.

Radical Neck Resection

Localized lesions with a suspected or proven cervical lymphatic involvement require a wide resection of the primary tumor within dissection of the involved neck. It involves excision of the cervical lymphatic system and removal of the sternocleidomastoid and omohyoid muscles, internal and external jugular veins, accessory nerve, submandibular gland and the inferior lobe of the parotid gland.

Commando Operation

It includes en bloc resection of primary tumor with the involved adjacent osseous structures and total radical neck dissection.

Palliative Therapy

In the tumors which are non-resectable, debulking procedure or deliberate subtotal removal is done. It provides palliation of acute symptom as pain relief, control of infection or alleviation of airway obstruction. The benefit of palliative therapy is temporary but quality of life can be improved for the patient.

Advantages of Surgical Treatment

- *During surgery*—immediate microscopic examination of the surgical margins can be performed during surgery, by the frozen technique, to ensure the adequacy of the excision. If the tissue shows microscopic involvement, the surgical margins should be widened.
- *Prosthesis management*—surgical defects can be satisfactory treated either by primary or secondary reconstructive procedures or by prosthesis rehabilitation.
- *Newly discovered lesion can be treated*—during surgery, newly discovered primary residual or recurrent tumor can be excised.
- *Adjunct treatment*—surgical treatment can be used as an adjunctive treatment in combination with other principal treatments like chemotherapy or radiotherapy.
- *Reduction of size of tumor before radiotherapy*—bulky tumors which have a central core of necrotic and hypoxic cells can be surgically reduced before radiotherapy, which is most effective of other treatment modalities like debridement of osteoradionecrosis following radiotherapy.

Reconstruction

Surgical Reconstruction

Reconstruction following resection of oral carcinoma is designed both, to repair the cosmetic defect and to reestablish the functions of the lost tissue. Conservation of lower border of mandible should be attempted, whenever feasible.

Mandible bone can be replaced by a variety of natural and alloplastic materials. Blocks of bone from hip or rib have commonly been used. Metal implants have also been used but suffer from problems of rejection and instability at the site where they abut the bone.

Nowadays, free bone graft secured by microvascular anastomosis has been used. In graft, bone remains viable and the graft may include periosteum, muscle and skin.

Following hemimandibulectomy, some patients are happy to tolerate a deformity. In these cases, direct soft tissue closure is feasible. Scar contraction, which will result in mandibular deviation, can be minimized by initial intermaxillary fixation and then interdental traction with training elastic bands.

In the maxilla, a split thickness skin graft is used to cover the defect and later, obturator replaces the lost tissue. Soft tissue loss may be repaired by a variety of methods using tissue of local, regional or distant origin.

Maxillofacial Prosthodontics

While surgeons can provide coverage of the defect following resection of oral carcinoma, the return of function such as speech and mastication and optimal cosmetic repair often rely on the skills of the prosthodontist. Two phase of prosthodontic care may be needed; the initial for the provision of surgical splints and after sufficient healing, definitive prosthetic appliance can be constructed and fitted.

Chemotherapy

It is described in Chapter 43: Anticancer Drugs.

New Therapies of Management of Oral Cancer

Over the last some years, there are many new methods for the management of oral cancer are available. These are as follows:

Photodynamic Therapy (PDT)

The first description of PDT was given in 1904 by Von Tappener and Jadlbauer.

- *Principle*—it involves selective uptake and retention of preadministered photosensitizer in a tumor. This is followed by irradiation with LASER of particular wavelength. This will lead to necrosis of tumor due to formation of singlet oxygen.
- *Photosensitizing agent*—the most commonly used agent is porphyrins. Porphyrins are a product of hemoglobin. Porphyrins which are used are photofrin, 5-aminolevulinic acid and foscan.
- *Technique*—
 - *Injection of photosensitizing agent*—porphyrins should be injected at dose of 2 mg/kg in the bloodstream.
 - *Absorption by tumors cells*—this is absorbed by all the cells of the body, but it remain more in contact with the tumors cells as compared to normal cells.
 - *Laser application*—after 48 hours of injection of porphyrins, tumor is activated by Laser of wavelength of 628-635 with an interval of 4-6 hours.
 - *Absorption of light by photosensitizer*—Laser light is absorbed by porphyrins with production of oxygen.
 - *Destruction of tumors cells*—this oxygen destroys cancer cells and damages the blood vessels. This will further reduce the blood supply to the tumors causing cell.
- *Indication*—it is more commonly used in superficial premalignant and malignant lesion of the oral cavity. It

is also used as adjunct therapy for poorly resected tumors.

- *Advantage*—it is cost effective, avoids systemic treatment, and applicable where surgery is not possible.
- *Disadvantage*—as there is limited penetration, it can cause tissue necrosis only to depth of 0.5-1 cm. It cannot treat metastasis.

Intralesional Chemotherapy

In this, drug is injected directly in the tumor. Drug remains concentrated in the tumor cells rather than spreading to the surrounding normal tissue. This will reduce the toxicity and side effect of the drug.

Intra-arterial Chemotherapy

In this, drug is injected into the arteries which are supplied to the tumor. This therapy can help to inhibit the tumors growth with high remission rate. There is also low toxicity reported.

Intensity Modulated Radiation Therapy (IMRT)

It is also called as 'tomotherapy'. IMRT delivers planned, specified doses of radiation therapy directly to cancer cells at the targeted site. This will spare the surrounding healthy tissue.

- *Principle*—in this, software is used to deliver a precise radiation beam, to particular area of tumor. This will decrease harmful doses to healthy tissue.
- *Component*—it consists of image acquisition software, treatment planning software, treatment simulator, platinum medical linear accelerator, dynamic multileaf collimator.
- *Advantage*—advantage includes reduced xerostomia, reduced risk of myopathy, and improved rate of tumor control.

Immunotherapy

It is designed to repair, stimulate and enhance the body's immune response. Previous effort with interferon, interleukins as immunomodulator got limited benefits. Nowadays, newer therapies like immunologic gene therapy and radioimmunotherapy are currently under trials.

- *Immunologic gene therapy*—in this, we are increased immunogenic potential of tumor cells.
- *Radioimmunotherapy*—also called as 'targeted radiotherapy'. In this, radionucleotides are linked to antibodies in order to deliver toxins directly to the tumor target. Most commonly used radionucleotides are Yttrium-90 and Iodine-131. Radioimmunotherapy is effective for treatment of lymphoma.

Gene Therapy

It is defined as introduction of functional genetic material into target cells to replace or supplement defective gene.

- *Principle*—precise amount of genetic material is transferred into each target cells allowing the expression of the gene product without causing toxicity. It targets dominantly on activated oncogenes on the cancer cells.
- *Gene addition therapy*—the role of p53, a tumor suppressor gene in cell regulation cycle and apoptosis is well established. Hence p53 gene transfer is safe and well tolerated.
- *Antisense RNA therapy*—gene expression can be inhibited by RNA. The antisense RNA can prevent activity of several known oncogenes. Such therapy can be directed towards cancer cells inhibiting the expression of oncogenes.
- *Suicide gene therapy*—it involves the introduction of gene into cell that enables a prodrug to be activated into an active cytotoxic drug. Most commonly used are herpes simplex virus thymidine kinase which selectively targets activity dividing cells.

Cancer Vaccines

Cancer vaccines include identifying unique cancer cell antigen, placing the gene for tumor antigen into viral vector and attaching an adjuvant to tumor molecules in order to stimulate an immune response against tumor cell. It is of two types:

- *Therapeutic vaccine*—it is used to treat cancer. It prevents further growth of existing cells and prevents recurrence. It includes *antigen/adjuvant vaccines* (specific protein fragments or peptides are used to stimulate immune response against tumor cells), *whole cell tumor vaccine* (this is taken from patient's own tumor, i.e. autologous or from other patient, i.e. allogenic) and *viral vector and DNA vaccines* (these are nucleic acid sequence of tumor antigen to produce cancer antigen proteins).
- *Preventive/prophylactic vaccines*—these are designed to target cancer causing viruses, thus preventing viral infection. Viral protein on the outside coat of cancer causing viruses is commonly used as an antigen to stimulate the immune system for preventive vaccine.

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Introduction

Ulceration of the oral mucous membrane is a common occurrence in patients and may challenge the diagnostic acumen of the clinician. When observed in mouth, all these lesions have a similar appearance. As texture of oral mucosa is thin, mucosa is easily traumatized and infected by foods and oral mucosa in their appearance is nonspecific in the oral cavity. The ulcer is a Greek word meaning 'wound' or 'sore'. It has focus of necrotic tissue on the surface and a destroyed overlying epithelium.

An ulcer consists of margins, edges, floor and base. A *margin* is the junction between normal epithelium and the ulcer; so it is the boundary of the ulcer. An *edge* is the junction between a margins and the floor of the ulcer. Activity is maximum at the margins and edges of the ulcer.

Five common types of ulcer edges are seen.

- *Undermined edge*—it is mostly seen in tuberculosis ulcer. The disease causing the ulcer spreads and destroys the subcutaneous tissue faster than it destroys the skin. The overhanging skin is friable reddish blue and unhealthy.
- *Punched out edge*—it is mostly seen in gummatous ulcer or in deep tropic ulcer. The edges drop down at right angle to the skin surface.
- *Sloping edge*—sloping edge is seen mostly in healing, traumatic or venous ulcer which is reddish purple in color and consists of new healthy epithelium.
- *Raised and pearly white, beaded edge*—it is a feature of rodent ulcer. This type of edge develops in invasive cellular disease and becomes necrotic in the center.
- *Rolled out (everted) edge*—it is a characteristic feature of malignant ulcer. This ulcer is caused by fast growing cellular disease, the growing portion at the edge of the ulcer heaps up and spills over the normal skin to produce an everted edge.

- *Floor* is the exposed surface of the ulcer. The floor is composed of connective tissue fibrin with polymorpho-nuclear leukocyte infiltration. One must understand the difference between the floor (exposed surface within the ulcer) and the base (on which the ulcer rest) which is better felt than seen.

Formation of Ulcer

There are many ways by which ulcer develops. The simplest and most frequent development is through the destruction due to trauma. But many ulcers develop by systemic illness and hence, their pathogenesis is different. These lesions are preceded by the accumulation of fluid in subepithelial regions with the consequent formation of small blisters (vesicles) or large blisters (bullae) (**Fig. 17-1**). The presence of fluid, thickness of the epithelium and proliferation of the blister predispose to rupture; thus resulting in ulcer formation, e.g. bullous form of erythema multiforme.

Oral ulcerations are preceded by intense accumulation of inflammatory cells in the subepithelial regions, clinically visible as macular or papular eruptions. They may break down with resultant ulcer formation, as in certain cases of recurrent ulcerative stomatitis. Rarely, ulcerative lesions are preceded by an accumulation of fluid entirely within the confines of the epithelium forming an intraepithelial vesicle or bulla, as observed in pemphigus.

The life cycle of an ulcer consists of three phases (**Fig. 17-2**):

- *Extension*—the floor is covered with exudate and sloughs while the base is indurated. The edge is sharply defined and discharge is purulent or even blood stained.
- *Transition*—it is occupied for preparation for healing. The floor becomes cleaner, the slough separates; indurations of base diminish and the discharge becomes more serous. Small reddish areas of granulation tissue appear at the floor and these unite until the whole surface is covered.

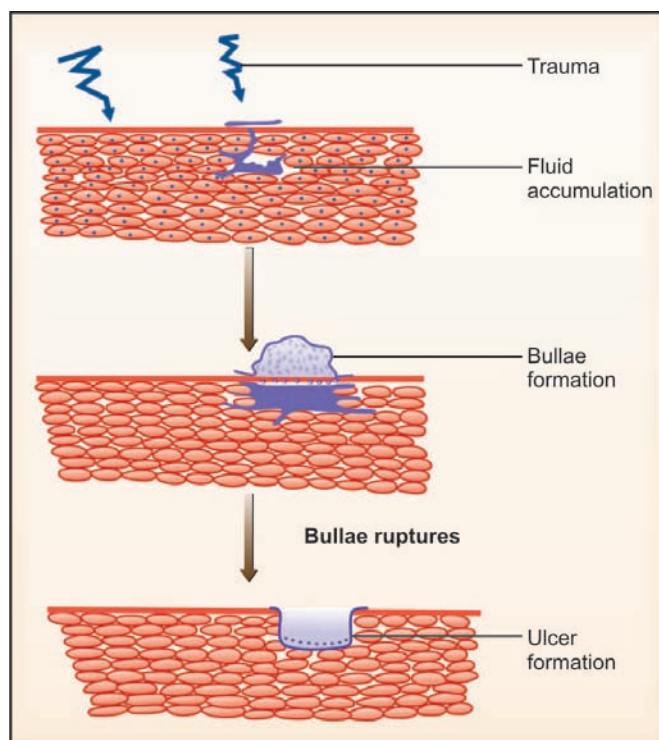


Fig. 17-1: Formation of ulcer (diagrammatic representation).

- **Repair**—it consists of the transformation of granulation tissue to fibrous which gradually contracts to form scar. The edge of an ulcer becomes more shelving as the epithelium gradually extends to cover the floor. This healing edge consists of three zones.
 - **Outer**—it consists of epithelium, which appears white.
 - **Middle**—it is bluish, where the granulation tissue is covered by few layers of epithelium.
 - **Inner**—inner reddish zone of granulation tissue covered by a single layer of epithelial cells.

Classification of Ulcers

1st Classification According to Etiology and Pathology

Traumatic

- Due to mechanical irritation
- Chemical irritant
- Thermal burns
- Radiation burns
- Anesthetics necrosis
- Oral trauma from sexual practice

Infection

Viral infection

- Herpes simplex infection

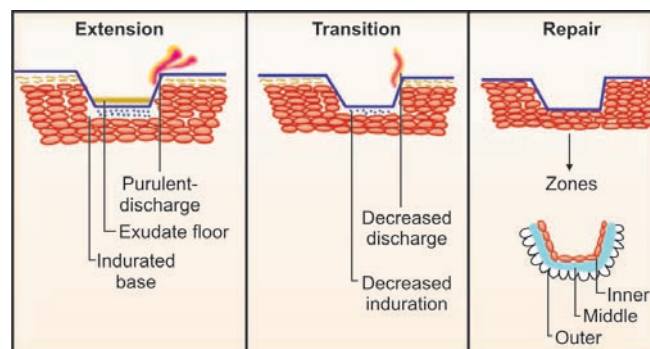


Fig. 17-2: Life cycle of ulcer (diagrammatic representation)

- Herpes zoster
- Hand-foot and mouth disease
- Herpangina
- Chickenpox
- Smallpox
- Measles
- Infectious mononucleosis
- AIDS—HIV

Bacterial infection

- ANUG
- Tuberculosis
- Syphilis
- Gonorrhea
- Scarlet fever
- Diphtheria

Fungal infection

- Histoplasmosis
- Blastomycosis
- Mucormycosis
- Cryptococcosis

Allergy

- Local (stomatitis venenata)
- Systemic
- Secondary vaccinia
- Acrodynia

Neoplastic

- Squamous cell carcinoma
- Malignant melanoma
- Non-Hodgkin's lymphoma

Systemic disorders

Blood disorder

- Agranulocytosis
- Cyclic neutropenia
- Leukemia
- Aplastic anemia

Nutritional deficiency

- Scurvy

- Riboflavin deficiency
- Pellagra
- Protein deficiency
- Malabsorption syndrome
- Xerostomia
- Hand-Schüller-Christian disease
- Letterer Siwe disease
- Acrodermatitis enteropathica (Zinc deficiency)

Disease of unknown etiology

- Aphthous stomatitis
- Erythema multiforme
- Pemphigus
- Bullous pemphigoid
- Mucous membrane pemphigoid
- Erosive lichen planus
- Epidermolysis bullosa
- Systemic lupus erythematosus
- Dermatitis sialometaplasia
- Wegener's granulomatosis
- Eosinophilic ulcer of the oral mucosa

Syndromes

- Stevens-Johnson syndrome
- Behcet's syndrome
- Reiter's syndrome

2nd Classification (Burkitt's)

Acute multiple ulcers

- Acute herpetic stomatitis
- Erythema multiforme
- Herpes zoster infection
- Cytomegalovirus infection
- Coxsackievirus infection
- ANUG
- Allergic reaction

Chronic ulcers

- Pemphigus
- Pemphigoid
- Cicatricial pemphigoid
- Epidermolysis bullosa
- Para-neoplastic pemphigus
- Linear IgA disease
- Subepithelial bullous dermatoses
- Chronic bullous disease of childhood

Recurrent ulcers

- Recurrent aphthous stomatitis
- Behcet's disease

Single ulcer

- Traumatic ulcer
- Eosinophilic ulcer of tongue
- Histoplasmosis

- Blastomycosis
- Mucormycosis

Ulcers Associated with Trauma

Traumatic Ulcer

It is a frequently encountered ulcerative lesion of mouth. The common term used to denote a traumatic ulcer is 'decubitus ulcer', 'tropic ulcer', 'neutrons-tropic' and 'Bednar's ulcer'.

Etiology

- *Mechanical or physical*—it includes biting, sharp or malposed teeth or roots, sharp food, stiff toothbrush bristles, sharp margins of crown, fillings, denture, orthodontic appliances and faulty instrumentation.
- *Self-inflicted*—this is also one of the important cause of traumatic ulceration.
- *Chemical*—it results from caustic substances such as silver nitrate, phenol, TCA, formocresol, eugenol, eucalyptus oil, phosphorus and acetylsalicylic acid.
- *Thermal*—excessive heat in the form of hot fluid or food, on rare occasion the application of the dry ice, reverse smoking and hot instrumentation.
- *Electrical current*—application to the oral tissues may result in destruction and consequent ulceration, e.g. galvanism.
- *Others*—radiation burns, self-inflicted and iatrogenic.

Clinical Features

- *Age and sex*—it occurs in persons of any age, with equal frequency in both the sexes.
- *Site*—it may involve any region of the mouth but common on tongue, in mucobuccal fold, gingiva and palate.
- *Factors affecting appearance of ulcer*—the appearance of the traumatic ulcer varies markedly depending on the site of the injury, the nature and severity of trauma and the degree of secondary infection present.
- *Uncomplicated ulcer*
 - *Symptoms*—there is tenderness and pain in the area of lesion and it will be helpful to identify the cause of lesion. It may persist for few days and may last for weeks.
 - *Size and shape*—the most common variety of traumatic ulcer is single uncomplicated ulcer. It is of moderate size (from several millimeters to a centimeter or more in diameter). Shape of ulcer is usually round, oval or elliptical in shape and flat or slightly depressed (Fig. 17-3).
 - *Surface*—its surface consists of a serosanguineous or grayish serofibrinous exudate. It may be composed of a grayish necrotic slough which when removed,

reveals a red raw tissue base. In case of ulcer on vermillion border lip, it may have crusted surface due to absence of saliva.

- *Margins*—margin of the lesion is surrounded by a narrow border of redness.
- *Crater-like ulcer*—a traumatic ulcer may be crater like generally due to the result of repetitive trauma over a prolonged period thereby preventing healing.



Fig. 17-3: Traumatic ulcer seen intraorally on lingual side which is oval in shape.

- *Complicated ulcer*—in still other instances, the traumatic lesions are large and irregular (**Fig. 17-4**). These are the result of unusually severe traumatic episodes, such as a blow or a fall and are often accompanied by considerable edema, inflammation and swelling of the neighboring tissues.
- *Infected ulcer*—the infected ulcer is larger, more irregular and more protruding than the noninfected one and often it is covered with a thick layer of necrotic slough through which purulent exudate may be observed. In case of infected ulcer, painful regional lymphadenopathy is also present.



Fig. 17-4: Large and irregular ulcer occur due to blunt trauma to the patient.

- *Progress*—usually the simple and uncomplicated traumatic ulcer heals uneventfully in 5 to 10 days after onset and even without treatment. However, in the presence of secondary infection or repetitive trauma, longer healing period is required.

Diagnosis

- *Clinical diagnosis*—there is adjacent source of irritation present. History of the patient is also aid in coming to diagnosis.

Differential Diagnosis

- *Primary syphilitic lesion*—painless indurated edema, painless lymph nodes swelling.
- *Tubercular ulcer*—undermined flabby borders usually painless.
- *Dystrophic ulcer*—rarely, ulceration is caused by deficient blood supply (pressure vasoconstriction) due to local anesthesia. Main localization is the hard palate. Explanation is obtained from patient's history.
- *Necrotizing sialometaplasia*—it is rare, and limited to palate area. In this case, there are nonpainful areas of necrosis.

Management

- *Removal of causative agent*—causative agent should eliminate.
- *Persistent ulcer*—triamcinolone acetonide in emollient base before bedtime and after meals. A persistent ulcer, not responding to the foregoing regimen, should be surgically excised and the entire tissue must be sent for histopathological examination to rule out dysplastic changes.
- *Pain relief*—dyclonine HCL or hydroxypropyl cellulose films can be applied for temporary pain relief.
- *Chlorhexidine gluconate* (0.2%) ointment, chlorhexidine mouthwash or even topical local anesthetic to relive acute symptoms of pain.

Anesthetic Necrosis

In some cases, after administration of local anesthetics ulceration and necrosis can occur at the site of injection.

Causes

- *Faulty technique*—in some cases, due to subperiosteal injection, necrosis can occur.
- *Administration of excess solution*—anesthetic necrosis can also occur in case of excess administration of solution in the area of tissue firmly bound to bone.

Clinical Features

- *Location*—it is most commonly seen in hard palate area.
- *Appearance*—well-circumscribed lesion develops at the site of injection.
- *Sequestration of bone*—in some cases, sequestration of bone may occur.
- *Healing*—healing is usually delayed in this case.

Diagnosis

- *Clinical diagnosis*—ulceration and necrosis at the site of injection will easily diagnose this condition.

Management

- It is usually not required and lesion will heal on its own.

Oral Trauma from Sexual Practice

Orogenital sexual practice can lead to ulceration in the oral cavity.

Etiology

- *Fellatio*—in this, there is extravasation of erythrocyte which results from soft palate elevation and tensing against environment of negative pressure.
- *Cunnilingus*—as tongue is moved forwards taut frenum rubs against the incisal surface of mandibular incisor.

Clinical Features

- *Location*—it is more commonly seen on soft palate (fellatio) and floor of mouth (cunnilingus).
- *Fellatio lesion*—in this, there is submucosal palatal hemorrhage. Lesion appears as erythema, petechiae, purpura or ecchymosis.
- *Cunnilingus*—horizontal ulceration with linear fibrous hyperplasia can be seen.

Diagnosis

- *Clinical diagnosis*—history of patient will give clue to the diagnosis.

Management

- No treatment is required in this condition and lesion is resolved after some days.

Ulcers Associated with Infection

It is described in respective chapters.

Ulcers Associated with Allergic Reaction

Antigen-antibody reaction can cause clinical diseases of mouth and face.

Drug Allergy

It is also called as 'drug idiosyncrasy', 'drug sensitivity' and 'stomatitis or dermatitis medicamentosa'. Some patients have greater susceptibility to drugs and manifest reactions more readily than others.

Drugs which can most commonly cause drug reactions are aminopyrine, barbiturates, gold, bromide, penicillin, streptomycin, opioid and morphine derivative, amphetamine, etc. Drug allergy many time is caused by drug abuse/misuse of the drugs. Many times, patient goes for self-medication causing various types of drug sensitivity reaction in the body. While diagnosing, drug abuse history of patient should be carefully noted. In the history, patient may give vague complaint or he may be polydrug user.

Clinical Features

- *Symptoms*—patient complains of fever, and arthralgia.
- *Signs*—it is characterized by inflammation, ulceration and vesicle formation. Lymph nodes enlargement is the common occurrence.
- *Skin lesion*—the skin lesion is often of erythematous type (**Fig. 17-5**), as in erythema multiforme. They may be urticarial in nature.



Fig. 17-5: Erythematous fixed drug reaction seen on the hand of patient (Courtesy Dr Sanjay Pincha).

- *Fixed drug reaction*—fixed drug reactions may occur in those who are administered on repeated occasions, a drug to which they are sensitive. It consists of appearance of same reaction at the same site at every time (**Fig. 17-6**).



Fig. 17-6: Fixed drug reaction

Oral Manifestations

- **Symptoms**—xerostomia is most common complaint in drug abuser. This may occur due to pharmacological action on the salivary gland resulting in the hypofunction of the gland. Another complaint includes taste alteration, eating difficulties.
- **Signs**—in the early stages of reaction, vesicle or even bullae may be found on the mucosa (Fig. 17-7). Occasionally purpuric spots appear and angioneurotic edema is seen.



Fig. 17-7: Vesicles seen on the buccal mucosa in the patient of drug reaction (Courtesy Dr Revent Chole).

- **Appearance**—the oral lesions are diffused in distribution and vary in appearance from multiple areas of erythema (Fig. 17-8) to extensive areas of erosion or ulceration (Fig. 17-9).
- **Gingiva**—ulceration and necrosis of gingiva often resemble ANUG.
- **Periodontal problems**—severe periodontal problems can be present with sloughing of epithelium.



Fig. 17-8: Extensive area of erythema seen on the palate due to drug allergy (Courtesy Dr Revent Chole).



Fig. 17-9: Extensive erosion of lip seen in the drug allergy (Courtesy Dr Amit Parate).

- **Teeth**—severe erosion of enamel of teeth may occur due to snorting (through) of drug. Snorting will result in bathing of teeth with corrosive substance as drugs can go back to the oropharynx through posterior nasopharynx.

Diagnosis

- **Clinical diagnosis**—medical history of the patient should be properly taken. Mucosal ulceration may be clearly diagnosed after caseation of drugs.
- **Laboratory diagnosis**—serum evaluation of generic antinuclear antibodies should be carried out.

Differential Diagnosis

- **Recurrent herpes simplex infection**—it has got virus based etiology, prodromal symptoms. Lesion occurs usually in cluster.

- *Chronic pemphigus*—age, systemic symptoms, skin findings, immunofluorescence.
- *ANUG*—systemic symptoms, punched out ulcerations.
- *Mucous membrane pemphigoid*—eye involvement and chronic in nature.
- *Erythema multiforme*—lip changes, fever and malaise. In severe conditions, target or iris type of skin lesions are seen.

Management

- *Discontinuation of drug*—the signs and symptoms of drug allergy regress with discontinuing of the causative drug.
- *Antihistaminic drugs*—the acute signs may be relieved by administration of anti-histaminic drugs.
- *Topical corticosteroid*—localized reaction can be resolved by using topical corticosteroid.
- *Management of anaphylactic stomatitis*—this is managed by administration of adrenaline to patient.

Contact Allergy

It is caused by delayed type of hypersensitivity reaction to topical antigen. On the skin, it is referred as '*dermatitis venenata*' and oral lesions are referred as '*stomatitis venenata*'. The oral mucosa is less sensitive as compared to skin as saliva dilutes and removes many antigens. Another reason is that period of contact with the oral mucosa is brief as allergen is rapidly dispersed from the oral cavity. Oral lesions are rare due to number of '*Langerhans cells*', saliva which dilutes the allergens and washes them from the surface of the mucosa and digest with enzymes and a thin layer of keratin present on the oral mucosa.

Etiology

- *Medication*—it is caused by poison ivy, leather, rubber, nickel, medication or other chemicals.
- *Dental amalgam*—contact allergy to dental amalgam is caused by mercury, which is released during condensation.
- *Dental and cosmetic preparation*—it includes dentifrices, mouthwashes, denture powder, lip stick, cough drop and chewing gums.
- *Dental material*—it includes vulcanite, acrylic, metal alloy base.
- *Dental therapeutic agents*—it includes alcohol, antibiotics, chloroform, iodide, phenol, procaine and volatile oils.
- *Cinnamon flavoring*—cinnamon oil is used as flavoring agents in ice cream, soft drinks, alcoholic beverage, gum, candy, mouthwashes, and breathes freshener.

Clinical Features

- *Age*—it has got more predilections of females as compared to males.

- *Skin lesions*—there are typically itching erythematous areas with superficial vesicle formation, directly at the site where allergen contacts skin. The skin may become thickened and dry.
- *Symptoms*—burning is very common complaint rather than itching of skin.
- *Signs*—after the rupture of vesicle, erosion may become extensive and if secondary infection occurs, the lesion may be serious.

Oral Manifestations

- *Acute contact stomatitis*—in this case, barely visible redness is present. Edema may be present. If vesicle present, it gets ruptured to form erosive lesion.
- *Chronic contact stomatitis*—in this, as allergen remains more in contact with the oral mucosa, we can see white hyperkeratotic lesion (**Fig. 17-10**).



Fig. 17-10: Stomatitis venenata showing white hyperkeratotic lesion on lip.

- *Plasma cell gingivitis*—another oral manifestation is plasma cell gingivitis, which is characterized by erythematous edematous attached gingiva accompanied by cheilitis and glossitis.
- *Toothpaste contact allergy*—this will cause widespread erythema with desquamation of superficial layer.
- *Cinnamon contact allergy*—allergy of cinnamon oil or formalin present in toothpaste appears clinically as swelling, cracking and fissuring of lips, perioral desquamation and edema, angular cheilitis, swelling of gingiva and oral ulcerations. In some cases, significant thickening occur and it may resembles hairy leukoplakia.
- *Dental amalgam contact allergy*—in this condition, most commonly affected area is posterior buccal mucosa and lateral border of tongue. It present as white or erythematous patch with or without peripheral striae. These lesions can be termed as lichenoid reaction to dental amalgam.

Diagnosis

- *Clinical diagnosis*—by taking proper history you can make out relationship between contact allergen and mucosal lesion.
- *Patch test*—suspected allergen is placed on normal non-hairy skin, i.e. on upper portion of back. It remains in contact with skin for 48 hours. Then the patch is removed and after 2 to 4 hours, the area is examined for persistent erythema.

Differential Diagnosis

- *Herpetic gingivostomatitis*—exanthemas on hand, feet and mouth.
- *Erythema multiforme*—lip changes, target or iris type of skin lesions and malaise, in severe conditions.
- *Acute necrotizing ulcerative gingivitis*—starting on tips of papillae, mild fever and lymphadenopathy. Punched out ulcerations covered with gray-yellow to gray-green pseudomembrane.

Management

- *Removal of allergen*—removal of suspected allergen is the treatment of choice in case contact stomatitis.
- *Antihistamine therapy*—in severe cases, antihistamine therapy combine with topical anesthetic give good response.
- *Topical steroid*—this is indicated in chronic cases. Most commonly used are fluocinonide gel and dexamethasone elixir.

Secondary Vaccinia

It is also called as '*vaccinia autonoculata*'. Undesired skin or mucosal lesion, after smallpox vaccination, is caused by transfer of the contents of vaccination pustule to other parts of the body (**Fig. 17-11**). It is followed by formation of secondary lesions usually with weaker reaction than the one seen in case of primary inoculation.



Fig. 17-11: Secondary vaccinia showing erythematous lesion around the lip which appears after vaccination.

Secondary vaccinia may develop at the site of scratching possibly in already existing epithelial defect. Eyes, ears and areas of lips and tongue are possible sites. In this area, a patch develops that becomes vesicular than pustular.

After the development of crust, lesion is repaired with scar formation. Other ulcerative lesions such as primary lesion of syphilis, tuberculosis, aphthae of the major type should be differentiated. The history is important as secondary vaccinia appear about 5 to 7 days after inoculation.

Acrodynia

It is described in Chapter 20: Oral Pigmentation.

Angioedema

It is also called as '*angioneurotic edema*', '*Quincke's edema*', and '*giant urticaria*'. It is common form of edema which involves subcutaneous and submucosal connective tissue.

Etiopathogenesis

- *Food allergy*—angioedema most commonly due to food allergy.
- *Physical stimuli*—some physical stimuli like heat, cold, exercise, emotional stress, and solar exposure can cause mast cell degranulation which in turn release histamine with resultant angioedema.
- *Angiotensin converting enzyme inhibitors*—these are most commonly used for hypertension. It includes captopril, enalapril and lisinopril. These drug cause angioedema due to increase level of bradykinin.
- *Hereditary*—in some cases, angioedema results from activation of complement pathway.
- *Mechanism*—the mechanism of development of swelling is due to vasodilation brought about by the release of histamine like substances with subsequent transudation of plasma.

Clinical Features

- *Age and sex*—it affects both sexes equally, but it is infrequent in children while some cases originate at puberty.
- *Sites*—most often, the face and lips are involved, but sometimes the tongue also becomes swollen.
- *Symptoms*—a feeling of tenderness or an itching or prickly sensation sometimes precedes the urticarial swelling.
- *Appearance*—it typically manifests as a smooth, diffuse edematous swelling, particularly involving the face, around the lips (**Fig. 17-12**), chin and eyes, the tongue and sometimes, the hands and feet.

- *Signs*—edema may develop gradually in a matter of hours, but can also progress in minutes.
- *Parotid gland*—parotid gland may be affected in some cases.
- *Eyes*—the eyes may be swollen, shut and lips may be extremely puffy.
- *Progress*—the condition usually last for 24 to 36 hours, although some cases persist for several days.
- *Hereditary form*—the hereditary forms are more dangerous because there is visceral involvement. Vomiting and abdominal pain may occur and especially, dangerous edema of glottis can result in death through suffocation.



Fig. 17-12: Diffuse edema seen around the lip of the patient in case of angioedema

Diagnosis

- *Clinical diagnosis*—clinical presentation is typical with known antigenic stimulant.

Management

- *Removal of cause*—when etiological agent such as food can be discovered, its elimination from diet will prevent recurrent attacks. If it is associated with ACE inhibitor, the drug should be avoided in future.
- *Antihistamine drugs*—antihistaminic drugs (50 mg to 75 mg diphenylhydramine hydrochloride) can give prompt relief. In case of severe attack, intramuscular epinephrine should be administered.

Ulcers Associated with Malignancy

It is described in Chapter 16: Malignant Tumor of Jaw.

Systemic Disorders

It is described in other respective chapters.

Diseases of Unknown (Uncertain) Etiology

Aphthous Stomatitis

It is a common disease characterized by development of painful, recurrent, solitary or multiple ulcerations of the oral mucosa, with no other signs of any other disease.

Etiology

- *Immunological abnormalities*—mucosal destruction occur due to T cell mediated immunological reaction. IgG and IgM binding of the epithelial cells of the spinous layer of oral mucosa is seen in patients suffering from recurrent aphthous ulcer.
- *Genetic predisposition*—increased susceptibility to RAS is seen among the children of RAS positive parents. Specific HLA antigen has been identified in RAS patients. There is familial tendency for the occurrence of the disease. It is associated with HLA histocompatibility agents.
- *Microbial organism*—a pleomorphic transitional L-form of α -hemolytic streptococcus and streptococcus sanguis has been implicated as the causative agent of the disease. The reason for their occurrence is increased in antigenic exposure of the patient. Other microbial agents which are responsible are varicella zoster virus, adenovirus and cytomegalovirus.
- *Systemic factors*—small percentage of patients with recurrent aphthae have certain nutritional deficiency. Presence of a deficiency allows the expression of an unrelated, underlying tendency to ulceration. Other systemic factors like celiac disease, cyclic neutropenia, inflammatory bowel disease, MAGIC syndrome (mouth and genital ulcer with inflamed cartilage) and Sweet syndrome can also cause aphthous ulcer in oral cavity.

Precipitating Factors

- *Trauma*—local trauma including self-inflicted bites, oral surgical procedures, toothbrushing, needle injections and dental trauma can cause aphthous ulceration. The reason behind is that it decreases the mucosal barrier locally.
- *Endocrine conditions*—there is some relation between occurrence of aphthous ulcer and pregnancy, menstruation and menopause. There is remission of ulcers during pregnancy. Incidences of aphthae are greatest during menstruation. Ulcerations are maximum during postovulation period.
- *Stress*—acute psychological problems appear many times, to precipitate the attacks of the disease. Stress and anxiety can also precipitate the attack. You can see more

aphthous ulceration during exam period and less ulceration in vacation.

- *Cessation of smoking*—it increases the frequency and severity of RAS.
- *Allergic factor*—patients may have a history of asthma, hay fever and food or drug allergy.

Classification

- *Minor aphthae*—it is also called as 'canker sores' in which the ulcers are less than 1 cm in diameter and heal without scar.
- *Major aphthae*—it is called as 'Sutton's disease' or 'peradenitis mucosa necrotica recurrens' and the ulcers are over 1 cm in diameter and heal with scarring.
- *Herpetiform ulcers*—recurrent crops of dozens of small ulcers throughout the oral mucosa.
- *Aphthous ulcer with Behcet's syndrome*—recurrent ulcers associated with Behcet's syndrome.

Clinical Features

- *Age and sex distribution*—it usually occurs between second and third decades of life. It is common in women than men.
- *Sites*—it occurs most commonly on buccal and labial mucosa (**Fig. 17-13**), buccal and lingual sulci, tongue, soft palate, pharynx and gingiva.



Fig. 17-13: Minor aphthous ulcer seen on labial mucosa in female patient.

- *Prodromal symptoms*—it begins with prodromal burning, itching or stinging for 24 to 48 hours, before the ulcer appears. Ulcer gradually enlarges over next 48 to 72 hours.
- *Symptoms*—the lesion is typically very painful so, it commonly interferes with eating for several days.
- *Signs*—it begins as a single or multiple superficial erosion covered by gray membrane. It is surrounded by localized areas of erythema (**Fig. 17-14**) and develops within hours.

- *Shape*—lesions are round, symmetric and shallow but no tissue tags are present from the ruptured vesicles.
- *Course of disease*—multiple lesions are present but the number and size are frequently varied. Most patients have between 2 to 6 lesions at each episode and experience several episodes a year. The ulcers themselves generally persist for 7 to 14 days.



Fig. 17-14: Aphthous ulcer surrounded by zone of erythema and covered by gray membrane.

- *Minor aphthae (Figs 17-15A and B)*—size is 0.3 to 1 cm. These ulcers heal without scarring, within 10 to 14 days.
- *Major aphthae (Sutton's disease)*—size of lesion is larger than 1 cm (**Fig. 17-16**) and may reach upto 5 cm in diameter (**Fig. 17-17**). They interfere with speech and eating. Large portions may be covered with deep painful ulcers. The lesions heal slowly and leave scars, which result in decreased mobility of uvula and tongue (**Fig. 17-18**) and destruction of portions of oral mucosa.
- *Herpetiform ulcers*—multiple small shallow ulcers often up to 100 in number. It is found on any intraoral mucosal surface. It begins as small pinhead size erosions that gradually enlarge and coalesce. Lesions are more painful than would be suspected by their size. It is present continuously for one to three years, with relatively short remission. Patient gets relief immediately with 2% tetracycline mouthwash.

Diagnosis

- *Clinical diagnosis*—diagnosis is made from clinical presentation and exclusion of other disease.

Differential Diagnosis

- *BMMP and pemphigus*—absence of vesicles and blebs.
- *Bednar's aphthae*—these are not aphthae as such but traumatic lesions on the palatal mucosa of a newborn which develop through careless wiping of the oral cavity.



Figs 17-15A and B: Minor aphthous ulceration seen on labial mucosa and gingiva. Size of ulcer is around 0.5 mm.



Fig. 17-16: Major aphthous ulcer seen on maxillary tuberosity area with size about 2 cm.

- *Cyclic neutropenia*—appearance with marked period of 3 weeks of compromised general condition, blood picture and granulocyte count should be taken into consideration.



Fig. 17-17: Sutton's disease measuring about 5 cm in diameter occurring on the soft palate (Courtesy Dr Soni).



Fig. 17-18: Major aphthous ulcer seen on the lateral border of tongue (Courtesy Dr Chole).

- *Erythema multiforme*—aphthous ulcers are uniform in distribution, crusted appearance with scaly crust of lips, target lesions and erosion of different sizes.
- *Necrotizing sialometaplasia*—rarely, limited to hard and soft palate, usually painless.
- *Lupus erythematosus and psoriasis*—white components are present.
- *Erosive lichen planus*—whitish changes in border lesion.
- *Primary syphilitic lesion*—painless, indurated edema, painless swollen lymph nodes.
- *Atrophic candidiasis*—smear test should be done for candida organism.
- *Herpetic stomatitis*—patient has fever, malaise and nausea with pinpoint size ulcers of gingiva.
- *Herpangina*—affects children in late summer and early winter and localized on soft palate.

- *Hand, foot and mouth disease*—vesiculo-ulcerative lesions occur simultaneously in the oral cavity and on hands and feet.
- *Recurrent herpes simplex infection*—difference between herpes simplex and aphthous ulcer are described in **Table 17-1**
- *Bite wound after local anesthesia*—it is described in **Table 17-2**.
- *Aphthae in Behcet's syndrome*—it is described in **Table 17-3**.
- *Squamous cell carcinoma*—it is described in **Table 17-4**.

Table 17-1: Difference between herpes simplex and aphthous ulcer

Features	Recurrent aphthous ulcer	Recurrent intraoral herpes infection
Age	Wide range	At any age, but more common in middle and older age.
Location	On freely movable mucosa like lip, buccal mucosa, tongue, mucobuccal fold and soft palate.	On buccal mucosa tightly bound to periosteum, hard palate, gingiva and alveolar ridge.
Initial lesion	Erythematous macule or papule which undergoes central blanching followed by necrosis and ulceration.	Clusters of small discrete gray or white vesicles without red erythematous halo, vesicle quickly ruptures forming small punctured ulcer of 1mm or less in diameter.
Mature lesion	Shallow ulcer, 0.5 to 2 cm in diameter, yellow necrotic center, regular border contrast, erythematous halo.	Shallow ulcer, larger than 0.5 cm lesions coalesce to form large lesions.
Number of lesion	Occur single	Usually several small punctate ulcers in clusters, regular border, round, variable erythematous halo.
Histology	Nonspecific ulcer	Epithelial cell with ballooning degeneration, multi-nucleated giant cell, virus particles

Table 17-2: Bite wound after local anesthesia

Chronic recurrent aphthae	Bite wound after local anesthesia
Roundish oval form	Irregular ragged margins
Erythematous halo	Inflammatory erythema is ill defined
Very painful	Moderately painful
Recurring appearance at different location	Lesion occurs only once

Table 17-3: Aphthae in Behcet's syndrome

Chronic recurrent aphthae	Aphthae in Behcet's syndrome
Aphthae is the only symptom	Mouth aphthae are part of a systemic disease
Superficial ulceration	Deep ulceration
Aphthae mostly in the anterior region of mouth	Aphthae also in the posterior region of mouth

Table 17-4: Squamous cell carcinoma

Aphthae (major type)	Squamous cell carcinoma
At any age	More than 50 years of age
History of quick development	Slow development
Very painful at onset, later moderately painful	Little or no pain
Inflamed border edges	Thickened edges
Lymph node often painfully swollen	Lymph nodes swollen without pain
Associated edema	Surroundings are not irritated
Palpation noncontributing	Borders indurated

Management

Minor aphthous or herpetiform aphthous

- *Topical corticosteroid*—topical corticosteroid triamcinolone acetonide 3 to 4 times daily. Other topical steroids which can be used are betamethasone syrup and 0.01% dexamethasone elixir. It should be used by *rinse and expectorate method*. In patient with localized ulceration, 0.05% betamethasone dipropionate or 0.05% fluocinonide can be used.
- *Anesthetic cream*—topical protective emollient base (Orabase) should be given.
- *Tetracycline mouth wash*—topical tetracycline mouth wash (250 mg per ml) use four times daily for 5 to 7 days produces good response in nearly 70% of the patients.
- *Nutritional supplement*—replacement therapy with vitamin B₁₂, ferritin, folate and iron is also indicated.
- *Maintenance of oral hygiene*—chlorhexidine mouthwash is also given to patient to maintain oral hygiene.

Severe cases

- *Injection of steroid*—Injection of corticosteroid directly in lesion should be given. Most commonly used are triamcinolone acetonide.
- *Topical application*—topical preparation like 0.05% clobetasol cream, fluocinolone gel is also used. Triamcinolone tablet can be dissolved directly over the lesion.
- *Beclomethasone dipropionate spray*—this is recommended in severe case of aphthous ulcer where it is difficult to reach. It is usually given in case of aphthous ulcer of tonsillar pillar.

- **Systemic steroid**—in resistant cases, systemic steroid can be given. Most commonly prednisone or betamethasone syrup in a *swish and swallow method* (patient gargle the syrup and then swallow it) is given. In some cases prednisone tablet is also given.
- **Chlortetracycline**—it is used as mouth rinse to be flushed over the affected region, for at least 2 minutes provides relief from pain.
- **Surgery**—surgical removal of aphthous ulcer can also be used. Laser ablation shortens the duration and decreases associated symptoms.
- **Other medication**—many other medication have been tried to resolve the disease. The success of the therapy is highly variable. The other therapies which can be used are dapsone, thalidomide, amlexanox, topical 5-aminosalicylic acid, azelastine hydrochloride, colchicines, cyclosporine, gamma globulin, hydrogen peroxide, pentoxifylline, prostaglandin E-2 gel, sucralfate, interferon alpha and levamisole can be used.

Erythema Multiforme

It is acute inflammatory disease of the skin and mucous membrane that causes a variety of skin lesions, hence the term 'multiforme'.

Etiology

- **Immune mediated disease** that is indicated by the deposition of immune complexes in the superficial microvasculature of the skin and mucous membrane or cell mediated immunity.
- **Drugs** like sulfonamides, trimethoprim, nitrofurantoin, phenylbutazone, digitalis, birth control pills and penicillin.
- **Microorganisms**—microorganisms like mycoplasma pneumoniae and herpes simplex virus.
- **Other factors**—vaccination, radiation therapy and occasionally other disease like Crohn's disease, ulcerative colitis and infectious mononucleosis can also predispose to this condition.

Types

- **Erythema multiforme minor**—it is self-limiting form and is less severe form and lesion.
- **Erythema multiforme major**—it is severe form may be present as Steven Johnson syndrome.
- **Toxic epidermal necrolysis**—it is the most severe form of erythema multiforme. It occurs due to increased apoptosis of the epithelial cells.

Clinical Features

- **Age and sex**—it is most frequently seen in children and young adults and is rare after the age of 50. It affects males more than females.

- **Sites**—most common area, involved are hands, feet, extensor surfaces of elbow and knees.
- **Onset**—it has got acute or explosive onset with generalized symptoms such as fever and malaise. It may be asymptomatic and in less than 24 hours, extensive lesions of oral mucosa may appear (**Fig. 17-19**).



Fig. 17-19: Large lesion seen on tongue which is showing bleeding tendency.

- **Appearance**—it is characterized by macule or papule, 0.5 to 2 cm in diameter, appearing in segmental distribution. Typical skin lesions of erythema multiforme may be non-specific macule, papule and vesicle.
- **Bull's eye or target lesion**—target or iris or bull's eye lesion consists of central bulla or pale clearing area, surrounded by edema and band of erythema.
- **Prognosis**—morbidity is high due to secondary infection, fluid and electrolyte imbalance or involvement of lungs liver and kidneys.
- **Stevens-Johnson syndrome**—in Stevens-Johnson syndrome, there is generalized vesicles and bullae formation involving skin, mouth, eyes and genitals.
- **Toxic epidermal necrolysis**—it is also called as 'Lyell's disease'. It occurs secondary to drug reaction and results in sloughing of skin and mucosa in large sheets. It is more common in female. It appears as patient is badly scalded. It is managed in burn centers where necrotic skin is removed under general anesthesia and healing takes place under sheets of porcine xenografts.

Oral Manifestation

- **Prevalence**—it occurs along with skin lesions in 45% of the cases.
- **Sites**—lip is prominently involved followed by buccal mucosa, palate, tongue and face.
- **Appearance**—oral lesions start as bullae, on an erythematous base and break rapidly into irregular ulcers.



Fig. 17-20: Extensive lesion of buccal mucosa seen in erythema multiforme



Figs 17-21A and B: Hemorrhagic crusting of lip seen in erythema multiforme patient (Courtesy Dr Chole).

- **Symptoms**—patient cannot eat or swallow and blood tinged saliva drools.
- **Signs**—the lesions are larger, irregular, deeper, and often bleed very freely. In full blown cases, lips are extensively involved (**Fig. 17-20**) and large portions of the oral mucosa are denuded of epithelium. Sloughing of mucosa and diffuse redness with bright red raw surface is seen.
- **Lips lesion**—lip lesion are extensive with presence of hemorrhagic crusting (**Figs 17-21A and B**).
- **Healing**—healing occurs in 2 weeks.

Diagnosis

- **Clinical diagnosis**—hemorrhagic crusting of lip with target lesion seen on skin will diagnose erythema multiforme.
- **Laboratory diagnosis**—the cutaneous or mucosal lesions generally exhibit intracellular edema of the spinous layer of epithelium and edema of superficial connective tissue, which may produce subepidermal vesicle. There is zone of liquefaction degeneration in the upper layer of the epithelium with intraepithelial vesicle formation and thinning, with frequent absence of the basement membrane.

Differential Diagnosis

- **Viral lesions like primary herpetic gingivo-stomatitis**—they are small, round, symmetrical and shallow but erythema multiforme lesions are larger, irregular, deeper, often bleed. Gingival involvement is rare in erythema multiforme.
- **Pemphigus vulgaris**—less associated inflammation, progressive course, immunological auto-antibodies against intercellular substance.
- **Mucous membranes pemphigoid**—slow development- eye changes, chronic course and frequent participation of gingiva.
- **Allergic reactions**—target lesions are absent.
- **Erosive lichen planus**—onset less acute, whitish lichen design on the border of erosion and skin changes.
- **Xerostomia**—mucosa is dry.
- **Herpetic gingivostomatitis** (**Table 17-5**).
- **Systemic lupus erythematosus** (**Table 17-6**).
- **Pemphigus** (**Table 17-7**).

Table 17-5: Herpetic gingivostomatitis

Erythema multiforme	Herpetic gingivostomatitis
Younger adults usually affected	Small children usually affected
Exudative component in the foreground: typical hemorrhagic crusty scales	Aphthous lesions over general stomatitis
Skin changes (target lesion)	No skin changes

Table 17-6: Systemic lupus erythematosus

Erythema multiforme	Systemic lupus erythematosus
Acute onset	Systemic disease with chronic course
Self-limiting course	Often lethal course
Skin changes (target lesion)	Polymorphous skin changes
Intraoral changes, especially in the anterior part of the oral cavity	Changes in the whole oral cavity with distinct erythematous component

Table 17-7: Pemphigus

<i>Erythema multiforme</i>	<i>Pemphigus</i>
Primarily in young adult	Usually in older patients
Acute disease	Chronic disease
Erosive crusty changes	Pustulopapular vegetation
Skin changes (target lesion)	Skin changes in intertrigous area
Negative Nikolsky's sign	Positive Nikolsky's sign

Diagnosis

- *Clinical diagnosis*—bull's eye lesion with hemorrhagic crusting of lip is typical of erythema multiforme.
- *Laboratory diagnosis*—biopsy shows subepithelial or intraepithelial vesiculation.

Management

- *Topical steroid*—mild cases can be managed by topical application of steroid.
- *Removal of cause*—if you are able to identify causative drug, its use should be discontinued.
- *Rehydration*—if patient is dehydrated due to pain then intravenous rehydration should be carried out. Patient should be given soft liquid diet.
- *Topical anesthetic mouthwash*—this is given to manage the pain in the oral cavity.
- *Systemic steroid*—in severe cases, systemic 30 mg/day prednisone or methyl prednisone for several days should be given. Dose should be tapered after the symptoms subside (**Fig. 17-22**).
- *Acyclovir*—it is indicated in erythema multiforme associated with HSV.
- *Management of toxic epidermal necrolysis*—this patient is managed in burn center. Administration of pooled human immunoglobulin is also effective in TEN. The reason behind is that it induced blockade of Fas ligand which induce epithelial cell apoptosis.



Fig. 17-22: Erythema multiforme lesion after week of steroid therapy (Courtesy Dr Bhaskar Patle).

Pemphigus

It is autoimmune disease involving the skin and mucosa and characterized by intraepidermal bulla formation.

Types

- *Pemphigus vulgaris*
- *Pemphigus vegetans*
- *Pemphigus foliaceus*
- *Pemphigus erythematosus*

Pemphigus Vulgaris

Mechanism

- *Epithelial cell separation*—binding of IgG antibody to Pemphigus antigen leads to epithelial cell separation by triggering complement activity or plasminogen plasmin system. Separation of cell takes place in lower layer of stratum spinosum.
- *Associated factors*—pemphigus may be associated with thymoma, myasthenia gravis or with multiple autoimmune disorders. It may be triggered by drug therapy like penicillamine, penicillin, phenobarbitone and captopril.

Clinical Features

- *Age and sex*—it is seen in 5th to 6th decades of life and male to female ratio is 1:1, with whites more commonly affected.
- *Size*—thin walled bullae or vesicles varying in diameter from few mm to several centimeters arise on normal skin or mucosa (**Fig. 17-23**).



Fig. 17-23: Thin walled bullae seen on back of patient (Courtesy Dr Bhaskar Patle).

- *Signs*—these lesions contain a thin, watery fluid shortly after the development, but this may soon become purulent or sanguineous. They rapidly break and continue to extend peripherally, eventually leaving large areas of denuded skin (**Fig. 17-24**).
- *Nikolsky's sign*—after giving application of pressure to an intact bulla, the bulla will enlarge by extension to



Fig. 17-24: Peripheral extension of vesicular lesion seen in pemphigus (Courtesy Dr Tapasya Karamore).

apparently normal surfaces. Another characteristic sign of the disease is that pressure to an apparently normal area will result in formation of new lesion. This phenomenon is called as Nikolsky's sign. It results from upper layer of skin pulling away from the basal layer. It is caused by prevesicular edema which disrupts the dermal-epidermal junctions.

- **Course**—the course of pemphigus vulgaris is a variable one; the disease terminating in death or recovery within a few days or weeks or it may get prolonged over a period of months or even years.

Oral Manifestations

- **Prevalence**—in 90% of the cases, oral lesions develop and in 60% cases, they occur first.
- **Sites**—initial lesion most frequently occurs on buccal mucosa because the epithelium demonstrates less intercellular substance and fewer intercellular junctions making the area more susceptible to acantholysis. Palate and gingiva are other common sites of involvement.
- **Onset**—oral lesions begin as classic bullae on non-inflamed base with formation of shallow ulcers as bullae break rapidly (**Fig. 17-25**).
- **Symptoms**—lesions bleed easily and are tender on palpation. The pain may be so severe that the patient is unable to eat.
- **Signs**—thin layer of epithelium peels away in an irregular pattern leaving denuded base.



Fig. 17-25: Ulcerative lesions seen on tip of tongue in pemphigus (Courtesy Dr Tapasya Karamore).

- **Nikolsky's phenomenon**—the oral lesions may exhibit Nikolsky's phenomenon and may be denuded by peripheral enlargement of the lesion (**Fig. 17-26**).
- **Margins**—the lesion may have ragged borders and be covered with white or blood tinged exudate. Edges of the lesion may extend peripherally.
- **Gingiva**—diffuse erythematous involvement of gingiva.



Fig. 17-26: Extensive lesion of oral mucosa in case of pemphigus showing denuded epithelium (Courtesy Dr Chole).

Pemphigus Vegetans

Types

- **Neumann type**—it is more common and early lesions are similar to those seen in pemphigus vulgaris.
- **Hallopeau type**—in hallopeau type; pustules, not bullae, are the initial lesions which are followed by verrucous hyperkeratotic vegetations.

Clinical Features

- **Incidence**—it occurs in 1% to 9% of the cases.

- *Appearance*—the flaccid bullae become eroded and forms ‘vegetations’ on some of the erosions.
- *Signs*—these fungoid masses become covered by purulent exudate and exhibit inflamed borders, frequently occur first on nose and in the mouth or axillae. The disease usually terminates in pemphigus vulgaris.

Oral Manifestations

- *Granular or cobblestone appearance*—gingival lesions are lace like ulcers with purulent surface on red base or have granular or cobblestone appearance.

Other Variant of Pemphigus

- *Pemphigus foliaceus*—it is a relatively mild form of pemphigus, which is most common in older adults. It is manifested by characteristic early bullous lesions which rapidly rupture and dry to leave masses of flakes on scales suggestive of an exfoliative dermatitis or eczema.
- *Brazilian pemphigus*—it is a mild endemic form of pemphigus foliaceus found in tropical regions, particularly in Brazil. It is seen in children and frequently in family groups.
- *Pemphigus erythematosus*—it is also called as *Senear-Usher syndrome*. It is form of disease which is characterized by the occurrence of bullae and vesicles concomitant with the appearance of crusted patches resembling seborrheic dermatitis or even lupus erythematosus. Most cases ultimately terminate in pemphigus vulgaris or foliaceus. The skin manifestations in any form of pemphigus may be accompanied by fever and malaise.
- *Paraneoplastic pemphigus*—it is associated with neoplasm like lymphoma or chronic lymphocytic leukemia. It occurs due to cross reactivity between antibodies produce by tumor and antigen associated with desmosomal complex. These multiple vesiculobullous lesion affects skin and oral mucosa. There is also palmar and planter bullae present which is not features of other type of pemphigus. In some cases, cicatrizing (scarring) conjunctivitis may develop.

Diagnosis

- *Clinical diagnosis*—positive Nikolsky’s sign with vesicular and bullous lesion seen on trunk, leg and oral mucosa.
- *Laboratory diagnosis*—basic defects are intraepithelial and is demonstrated as acantholysis as in *stratum spinosum*.
- *Tzanck smear*—it is done to demonstrate Tzanck cells which often are found lying freely within the vesicular space.
- *Indirect immunofluorescent antibody test*—antibodies against intercellular substance can be seen. The titers of

antibody are directly related to the level of the clinical disease.

- *Direct test*—antibody will bind the immunoglobulin deposit in the intercellular substance and show positive fluorescence under fluorescence microscope.

Differential Diagnosis

- *Recurrent aphthous stomatitis*—severe but heals and recurs; but in Pemphigus lesion, course extends over a period of weeks or months. Lesions of Pemphigus are not round, symmetrical but are shallow and irregular and often have detached epithelium at the periphery.
- *Dermatitis herpetiformis*—mucosal involvement is rare, Tzanck test and Nikolsky’s sign are negative. Skin changes are polymorphic.
- *Viral infection*—bullae are larger.
- *Bullous pemphigoid*—usually not intraoral; immunofluorescence can be done.
- *Erythema multiforme*—polymorphous appearance, systemic symptoms, acute onset, older patients.
- *Bullous drug induced exanthema*—negative-Tzanck test and Nikolsky’s sign, the vegetating variant can be confused with syphilitic patches and with other vegetating lesions caused by bromides, iodides, with acanthosis nigricans (pigmentation) and angular cheilitis.
- *Pyostomatitis vegetans*—pustule formation.
- *Wegner’s granulomatosis*—involvement of the nasal cavity, histology.
- *Benign mucous membrane pemphigoid* (Table 17-8).

Table 17-8: Benign mucous membrane pemphigoid

Pemphigus	Mucous membrane pemphigoid
Blisters filled initially with clear, and later with yellowish cloudy fluid.	Blisters do not get cloudy, change into erosions.
Pressure over blister causes it to shift laterally into adjacent mucosa.	Lateral shifting of the blisters usually not possible.
Skin changes.	Skin involvement less frequent.
No eye symptoms.	Eye involvement in 80% of the cases.
Severe illness picture.	No influence on general condition.
Painful lesions.	Blisters without symptoms.

Management

- *Corticosteroids*—topical or systemic prednisone. Systemic prednisone is used to bring disease under control and once the disease is under control, dose of prednisone is reduced.
- *Combination therapy*—high dose of corticosteroids are combined with immuno-suppressive drugs such as cyclosporine or azathioprine.

- *Plasmapheresis*—it is useful in patients refractory to corticosteroids.
- *Others*—parenteral gold therapy, etretinate and dapsone can also be used.
- *Newer therapy*—administration of 8-methoxypsoralen, followed by exposure of peripheral blood to ultraviolet radiation.

Bullous Pemphigoid

It is also called as '*para-pemphigus*', or '*aging pemphigus*'. In this, the initial defect is subepithelial in the lamina lucida region of the basement membrane. It is associated with anti-basement membrane antibodies which are detected in the basement membrane.

Clinical Features

- *Age and sex distribution*—it occurs chiefly in adults over the age of 60 is self-limiting and rarely lasts over 5 years.
- *Onset*—pruritus is seen in patient which is followed by develop of multiple bullae. Bullae do not extend peripherally and remain localized; heal spontaneously.
- *Skin lesion*—skin lesions begin as generalized non-specific rash, commonly on the limbs, which appear as blisters on inflamed skin; itching precedes.
- *Progress*—it may persist for several weeks to several months before ultimate appearance of vesiculo-bullous lesions.
- *Signs*—these vesicle and bullae are relatively thick walled and may remain intact for several days (**Fig. 17-27**). Rupture does not occur although it leaves raw eroded areas which heal rapidly.



Fig. 17-27: Intact bulla seen on hand of patient in bullous pemphigoid.

Oral Manifestations

- *Sites*—vesicles and ultimately erosion may develop not only on the gingival tissue but any other area such as the buccal mucosa, palate (**Fig. 17-28**), floor of the mouth and tongue.

- *Symptoms*—oral lesions are smaller, form more slowly and are less painful.
- *Gingiva*—gingival lesions consist of generalized edema, inflammation, desquamation, and localized areas of discrete vesicle formation.



Fig. 17-28: Ruptured bulla seen on palate of the patient in bullous pemphigoid (Courtesy Dr Soni).

Diagnosis

- *Clinical diagnosis*—bullae present on skin which does not extend peripherally. Ruptured bulla can be seen.
- *Laboratory diagnosis*—the vesicle and bullae are subepidermal and non-specific. Epithelium appears normal. The vesicle contain fibrinous exudate admixed with occasional inflammatory cells.
- *Indirect immunofluorescence antibody test*—lesions will demonstrate circulating IgG antibodies against basement membrane antigen.
- *Tzanck smear* negative to acantholytic cells.
- *Direct immunofluorescence testing and complement fixation test*—positive specimen will demonstrate IgG and complement in the basement membrane zone.

Management

- *Systemic steroids*—moderate doses of prednisone can be given. Later on, prednisone should be given in alternate day therapy.
- *Immunosuppressive therapy*—immunosuppressive drug like azathioprine may be given in this patient.
- *Other therapy*—other therapy like dapsone, tetracycline, and niacinamide can be given in this patient.

Benign Mucous Membrane Pemphigoid

It is also called as '*cicatricial pemphigoid*'. It is a disease of unknown etiology but probably is autoimmune in nature. In this condition, tissue bounded autoantibodies are

directed against basement membrane. The word cicatricial is derived from the word *cicatrix* meaning scar.

Clinical Features

- *Age and sex*—it occurs more commonly in patients over 50 years of age and female to male ratio is 2:1.
- *Sites*—typically, the vesiculo-bullous lesions occur on the oral mucus membrane, conjunctivae and skin. The other mucous membranes involved are those of nose, larynx, pharynx, esophagus, vulva, vagina and penis.
- *Symptoms*—involvement of esophagus and trachea may cause strictures leading to difficulty in swallowing or breathing.
- *Eyes*—adhesions may develop between bulbar and palpebral conjunctivae resulting in obliteration of the palpebral fissure with opacity of the cornea, frequently leading to blindness. Scarring can lead eyelid to turn inward (*entropion*). This will cause eyelashes to rub against cornea and globe (*trichiasis*).
- *Healing*—it may lead to scarring of affected area.



Fig. 17-29: Erosion seen on buccal mucosa of patient in benign mucous membrane pemphigoid (Courtesy Dr Chole).

Oral Manifestations

- *Sites*—it occurs on gingiva, buccal mucosa and palate. The mouth may be the only site involved.
- *Onset*—the mucosal lesions are also vesiculo-bullous in nature, but appear to be relatively thick walled and for this reason may persist for 24 to 48 hours before rupturing and desquamation.
- *Appearance*—there may be formation of ulcer, which surrounded by zone of erythema. There may be erosion on cheek (Fig. 17-29) and vesicles on palate and narrower peripheral extensions.
- *Signs*—after rupture of vesicle surface epithelium is lost leaving raw red bleeding surface (Fig. 17-30).

- *Desquamative gingivitis*—gingiva is edematous and bright red, involvement is patchy and diffuse.
- *Healing*—the oral lesions rarely produce scars.



Fig. 17-30: Same patient showing ulceration on inner aspect of lip.

Diagnosis

- *Clinical diagnosis*—desquamative gingivitis with erosive lesion seen in oral cavity with typical eye lesion will diagnose these conditions.
- *Laboratory diagnosis*—the vesicle and bullae are sub-epidermal rather suprabasilar and there is no evidence of acantholysis. Low titers of serum antibody.
- *Direct immunofluorescent study*—it will show positive fluorescence for immunoglobulins and complement in basement membrane zone, i.e. in intercellular substance of prickle cell layer of epithelium.

Differential Diagnosis

- *Pemphigus vulgaris*—painful erosions, eye involvement only in more severe cases, rapid appearance of blister, rapid transformation into erosion, thinner walled larger erosions.
- *Bullous pemphigoid*—primary site is skin.
- *Erythema multiforme*—acute onset, feeling of malaise, target lesions polymorphous picture and often self-limiting.
- *Behcet's syndrome*—erosive lesions, no bullae, no desquamative gingivitis and genital lesions.
- *Herpetic gingivostomatitis* (Table 17-9).

Management

- *Topical steroid treatment*—disease can be controlled by topical application of steroid many times daily. In case of gingival lesion, flexible mouth guard may be fabricated to use as carrier for the corticosteroid medication.
- *Systemic steroid and immunosuppressive therapy*—if topical treatment is not successful, systemic corticosteroid with

Table 17-9: Herpetic gingivostomatitis

Mucous membrane pemphigoid	Herpetic gingivostomatitis
Development of bullous erosive lesion on apparently healthy mucosa	Prodromal –feeling of tension, burning, vesiculo-erosive merging lesion
Can appear on any region of the mucosa	Almost only on the hard palate and gingiva
Usually in older patients	Independent of age
Eye changes are frequent	No eye involvement
Epithelial rest is at border	No epithelial rest at borders
Chronic course with intermittent bouts	Long asymptomatic interval between recurrence

immunosuppressive agents like cyclophosphamide may be used.

- *Dapsone therapy*—this sulfa drug derivative can be used as an alternative therapy. It should not be used in glucose 6 phosphate dehydrogenase deficiency.
- *Tetracycline and nicotinamide therapy*—another alternative therapy of tetracycline or minocycline and niacinamide can be given. It should be given in divided doses of 0.5 to 2 gm each.
- *Ophthalmologist consultation*—patient should be send to ophthalmologist for eye lesion.

Familial Benign Chronic Pemphigus

It is also called as *Hailey-Hailey* disease. It is uncommon disease transmitted by autosomal dominant gene. Heat and sweating amplify the outbreak of the lesion while spontaneous remission may occur in cold weather.

Clinical Features

- *Age and sex*—it is usually manifested during adolescent or young life with no predilection for sex.
- *Sites*—the most common sites are flexure surfaces of the axillae and groin, the neck and the genital area.
- *Appearance*—the lesions develop as small groups of vesicles appearing on normal or erythematous skin, which soon rupture to leave eroded, crusted areas
- *Nikolsky's sign*—they enlarge peripherally but heal in center with Nikolsky's sign positive.
- *Symptoms*—tender and enlarged regional lymph nodes may also be present.

Oral Manifestations

- *Eroded area*—intraorally, there is presence of crops of vesicle which ruptures very rupturing leaving raw eroded areas.

Diagnosis

- *Clinical diagnosis*—crops of vesicle seen on flexor surface of hand with positive Nikolsky's sign.
- *Laboratory diagnosis*—acantholysis is more extensive as compared to other types of pemphigus. Characteristic feature of this disease in occasional intercellular bridges persist so that adjacent epithelial cells still adhere to each other. This gives classic description of the 'dilapidated brick wall' effect.

Management

- *Antibiotics therapy*—it is generally effective in this lesion. Topical antibiotics like clindamycin or erythromycin in improvement of lesion.
- *Soothing ointment*—soothing ointment like aluminum acetate will give some relief from symptoms.
- *Corticosteroids*—this should be used intermittently in this disease.

Epidermolysis Bullosa

It is a dermatological disorder in which bullae or vesicles occur on skin or mucous membrane surface spontaneously, shortly after minor trauma. There is defect in attachment mechanism of epithelial cells.

Classification

- *Epidermolysis bullosa simplex*
 - Generalized form
 - Localized form (Weber-Cockayne syndrome, recurrent bullous eruption of hands and feet)
- *Epidermolysis bullosa dystrophic, dominant.*
- *Epidermolysis bullosa dystrophic, recessive.*
- *Junctional epidermolysis bullosa* (Epidermolysis bullosa latalis, junctional bullous epidermatosis, Herlitz's disease)—it is severe form and many patient may die at birth.
- *Epidermolysis bullosa acqvista (acquired)*—it is similar to dystrophic form of the disease but usually with an adult onset. It is autoimmune rather than genetic in origin.

Clinical Features

Epidermolysis bullosa simplex

- *Age*—it is inherited as autosomal dominant trait and manifests at birth or shortly thereafter.
- *Sites*—it is characterized by formation of bullae or vesicle on the hands and feet at site of friction or trauma. The knees, elbows and trunk are rarely involved and nails are occasionally affected.
- *Healing*—when the blister heals within 2 to 10 days there is no resultant scarring or permanent pigmentation.

- *Prognosis*—the disease appears to improve at puberty and prognosis is good for normal life span.
- *Localized form*—the localized form is limited to hands and feet only and tends to exacerbate in hot weather.

Epidermolysis bullosa dystrophica dominant

- *Age*—the onset is at infancy and it may delay until puberty.
- *Sites*—the blister commonly develop on the ankles, knees, elbows, feet and head.
- *Appearance*—the initial lesions are vesicle or bullae. Healing results in scarring which is sometimes keloid in type.
- *Signs*—hair may be sparse (**Fig. 17-31**), while nails are usually dystrophic (**Fig. 17-32**) or absent with milia present.
- *Palmar-planter keratoderma* with hyperhidrosis also may occur with ichthyosis and sometimes hypertrichosis. In some cases finger nail may be lost.



Fig. 17-31: Sparse hair seen in epidermolysis bullosa patient.



Fig. 17-32: Dystrophic nails seen in epidermolysis bullosa.

Epidermolysis bullosa dystrophica recessive

- *Age*—it has onset at birth or very shortly thereafter.
- *Appearance*—it is characterized by formation of bullae spontaneously or at sites of trauma, friction or pressure (**Fig. 17-33**).
- *Sites*—the typical sites of involvement are the feet, buttock, scapula, elbows, finger and occiput.



Fig. 17-33: Vesicular lesion seen on thigh of patient in epidermolysis bullosa

- *Nikolsky's sign*—Nikolsky's sign is positive in this type of epidermolysis bullosa.
- *Aspiration*—the bullae contain a clear, bacteriologically sterile or sometime blood tinged fluid.
- *Signs*—when these bullae rupture or are peeled off under trauma or pressure, they leave raw, painful surface.
- *Healing*—the bullae heal by scar and milia formation which may result in a functional club-like fists.
- *Hair and nails*—the hair may be sparse, while the nails are usually dystrophic.
- *Finger*—there may be fusion of the finger resulting in mitten like deformity.

Junctional Epidermolysis Bullosa

It is extremely severe form of the dystrophic recessive form, which is incompatible with prolonged survival. It has onset at birth, absence of scarring, milia, pigmentation and death within three months of age. The bullae are similar to the recessive form but they develop simultaneously and sheets of skin may actually be shed.

Oral Manifestations

- *Epidermolysis bullosa simplex*—bullae of the oral cavity are reported in occasional cases of generalized epidermolysis bullosa, but teeth are not affected.
- *Epidermolysis bullosa dystrophica dominant*—bullae can occur in this type sometimes, oral milia can be seen but

teeth are unaffected. There is also gingival recession and reduction in depth of buccal vestibule.

- *Epidermolysis bullosa dystrophic recessive*
 - *Prodromal signs*—they may be preceded by the appearance of white spots or patches on the oral mucous membrane or by development of localized areas of inflammation. Vesicle and bullae formation may be induced by food having some degree of texture.
 - *Symptoms*—these bullae are painful especially when they rupture or when the epithelium desquamates (**Fig. 17-34**). Scar formation results in obliteration of sulci and restriction of the tongue movement. Hoarseness and dysphagia may occur as a result of bullae of larynx and pharynx.
 - *Signs*—esophageal involvement produces serious strictures.
 - *Teeth*—dental defects like rudimentary teeth, congenitally absent teeth, hypoplastic teeth and crowns denuded of enamel may be seen.



Fig. 17-34: Rupture bullae seen on upper lip in epidermolysis bullosa patient

- *Junctional epidermolysis bullosa*—oral bullae are frequently very extensive and because of their extreme fragility produce serious feeding problems. Severe disturbance in enamel and dentin formation of deciduous teeth also occur.

Diagnosis

- *Clinical diagnosis*—Nikolsky's sign is positive with vesicle and bullous formation on skin, leg, thigh and face. The diagnosis should be simple when the family history, reaction to trauma and the lesions appearing most profusely on the extremities are taken into consideration.
- *Laboratory diagnosis*—individual cells become edematous and show dissolution of organelles and tonofibrils with displacement of the nucleus to the upper end of cells.

Management

- *Sterile drainage*—large blisters should be pricked and the blister fluid released. Dressing, to minimize reaction may be helpful.
- *Antibiotics*—super infections should be treated with appropriate local or systemic antibiotics.
- *Plastic surgery*—mitten deformity of hand can be corrected with plastic surgery.
- *Lip lubricant*—this should be applied to prevent trauma to lip.

Dermatitis Herpetiformis

It is also called as 'Dühring-Brocq disease'. It is a rare, benign, chronic, recurrent dermatologic disease of unknown etiology.

Clinical Features

- *Age and sex*—it occurs between 20 to 55 years of age, with males affected at least twice as frequently as females.
- *Sites*—these occur most frequently on buttocks, extremities as well as on the face, scalp and sometimes, the oral cavity.
- *Symptoms and signs*—the first manifestation of the disease is usually pruritis and severe burning followed by the development of erythematous papules (**Fig. 17-35**), vesicles, bullae or pustules. The patient usually shows increased severity in summer months.



Fig. 17-35: Papular lesion seen on leg of patient in dermatitis herpetiform (Courtesy Dr Sanjay Pincha).

Oral Manifestations

- *Ulceration*—vesicles and bullae rupture very rapidly in oral cavity. This rupture of vesicle will result in formation of superficial ulceration.

Diagnosis

- *Clinical diagnosis*—ulcerative lesion in oral cavity in association with pruritis lesion present on extremities will give clue to the diagnosis.
- *Laboratory diagnosis*—the lesion shows accumulation of neutrophils and eosinophils in the dermal papillae. This will result in microabscess formation. Direct immunofluorescence staining is positive at the epidermal-dermal junction. Patient develops eosinophilia and sensitivity to halogens (chlorine, bromine, iodine and fluorine), both by patch test.

Differential Diagnosis

- *Pemphigus*—chronic course, epithelial tag on the borders of lesion, occurs in older patients.
- *Mucous membrane pemphigoid*—erosion, immunofluorescence, no necrosis, eye involvement, Tzanck test is negative.
- *Erythema multiforme*—acute onset, most commonly seen on the mucosa of lip, iris or target lesions, more frequent in young patients.

Management

- *Dapsone*—100-200 mg per day will give prompt relief.
- *Gluten free diet*—a gluten free diet may help to reduce or completely remove the disease.

Eosinophilic Ulcer of the Oral Mucosa

It is described in Chapter 22: Diseases of Tongue.

Angina Bullosa Hemorrhagica

It is the term used to describe benign and generally sub-epithelial oral mucosal blister filled with blood. Initially, it is called as *traumatic oral hemophlyctenosis*. The term angina bullosa hemorrhagica is given by Badham in 1967.

Etiology

- *Mild form of epidermolysis bullosa*—it has been suggested that it could represent a mild localized form of epidermolysis bullosa.
- *Minor trauma*—it can occur due to minor trauma due to hot, crispy and coarse food. Other factors which can cause minor trauma are periodontal therapy, dental injection of anesthetics.
- *Long-term steroid inhaler therapy*—in some cases, it is associated with long-term steroid inhaler therapy.

Clinical Features

- *Age and sex distribution*—it affects middle age or elderly people with no sex predilection.

- *Location*—it usually occurs on the soft palate.
- *Onset*—it is characterized by the rapid appearance of solitary blood filled blister (hemorrhagic bulla).
- *Symptoms*—patient may complain of apparent tightness (angina) in the area immediately before and during the formation of swelling.
- *Signs*—bulla may spontaneously discharge to leave area of erosion. It heals without scarring. Size may increase up to 1-3 cm in diameter.
- *Respiratory obstruction*—as this condition is acute in some cases, it may cause acute respiratory obstruction to the patient.

Diagnosis

- *Clinical diagnosis*—occurrence of hemorrhagic bulla with complaint of tightness will give clue to diagnosis.

Management

Patient should be given antiseptic mouthwash to control infection. As blister spontaneously ruptures and heals rapidly, no other treatment is required.

Common Syndromes

Stevens-Johnson Syndrome

It is the severe form of erythema multiforme with widespread involvement, typically involving skin, oral cavity, eyes and genitalia.

Clinical Features

- *Symptoms*—it commences with abrupt occurrence of fever, malaise, photophobia and eruptions on oral mucosa, genital mucosa and skin.
- *Cutaneous lesions*—cutaneous lesions are similar to erythema multiforme and are hemorrhagic, often vesicular or bullous.
- *Eye lesions*—it consists of photophobia, conjunctivitis, corneal ulcerations. Keratoconjunctivitis sicca has also been described and blindness may result, chiefly from recurrent bacterial infection.
- *Genital lesions*—genital lesions are reported to consist of non-specific urethritis, balanitis and vaginal ulcers.
- *Complications*—other reported complications are related to respiratory tract involvement such as tracheo-bronchial ulcerations and pneumonia.

Diagnosis

- *Clinical diagnosis*—erythema multiforme with skin, eye and genital lesion will aid in diagnosis.



Fig. 17-36: Stevens-Johnson syndrome showing extensive lesion on the soft palate.

Oral Manifestations

- *Symptoms*—oral mucous membrane lesions may be extremely painful and so, mastication becomes impossible.
- *Signs*—mucosal vesicles or bullae occur, rupture and leave a surface covered with thick, white or yellow exudates (**Fig. 17-36**).
- *Pharynx*—erosions of the pharynx are also common.
- *Lips*—lips may exhibit ulcerations with bloody crusting and are often painful.

Management

- There is no specific treatment for this, although in some cases, ACTH, cortisone and chlortetracycline have shown promising results.

Behcet's Syndrome

It is discovered by H. Behcet in 1937 as a triad of recurring oral ulcers, recurring genital ulcer and eye lesion.

Etiology

- *Immunological*—it is caused by immune complexes that lead to vasculitis of small and medium sized blood vessels. It has got strong association with certain HLA type. There may be inflammation of the epithelium caused by immunocompetent T-lymphocytes and plasma cells.
- *Environmental antigen*—it may be associated with environmental antigen such as bacteria (streptococci), viruses, pesticides and heavy metal.

Classification

- *Mucocutaneous*—oral, genital and skin lesions.
- *Arthritic*—arthritis, in addition to mucocutaneous lesions.

- *Neuro-ocular*—neurologic, ocular and mucocutaneous lesions.

Clinical Features

- *Age and sex*—it begins between 10 to 45 years of age, with a mean age of occurrence of 30 years. It is five to ten times more common in males.
- *Recurring oral ulcers*—it may be mild or may be deep, large scarring lesions and may appear anywhere on the oral and pharyngeal mucosa. They are painful lesions. They may range from several millimeters to a centimeter in diameter. These ulcers have erythematous borders and are covered by gray or yellow exudate.
- *Recurring genital lesions*—it includes ulcers of scrotum and penis in males and ulcers of labia in females. The genital ulcers are small and painful in females.
- *Eye lesions*—consist of uveitis, retinal internal edema and vascular occlusion, optic atrophy, conjunctivitis and keratitis.
- *Skin lesions*—these are generally small pustules or papules on the trunk or limbs and around the genital.
- *Arthritis*—arthritis is common. Affected joint is red and swollen.
- *CNS involvement*—it may occur and it may include brain stem involvement of cranial nerve and neurologic degeneration.
- *Others*—thrombophlebitis, intestinal ulceration, venous thrombosis, renal and pulmonary disease.

Diagnosis

- *Clinical diagnosis*—recurrent oral ulcerations at least 3 times in 12 month period, plus at least two of the following four manifestations:
 - Recurrent genital lesions.
 - Eye lesions including uveitis or retinal vasculitis.
 - Skin lesions including erythema nodosum, pseudofolliculitis and papulo-pustular lesions.
- *Laboratory diagnosis*—there is positive *Pathergy test* (cutaneous hypertrophy to intra-cutaneous injection or needle sticks (Pathergy) with the finding of pustule forming 24 hours after needle puncture).

Management

- *Topical and intralesional steroids*—oral mucosal lesions be treated with topical or intralesional steroids.
- *Severe cases*—patients with life threatening or slight threatening vasculitis are managed with combination of immunosuppressive drugs and systemic corticosteroids. Immunosuppressive drugs which used are cyclosporine, azathioprine and interferon alpha. Use of

above therapy will result in decreases in ocular and systemic manifestation.

- *Cyclosporine and colchicines combination*—cyclosporine and colchicine in combination with corticosteroids have also been shown to be useful in mucocutaneous and gastrointestinal manifestations.
- *Plasmapheresis*—this is used in emergency.

Reiter's Syndrome

It is a disease of unknown etiology and is considered as an important complication of non-gonococcal urethritis and is often acquired sexually. Reiter syndrome can also be found in HIV infected patient. Oral lesions occur in less than 5% to about 50% of the patients with the disease.

It consists of tetrad of:

- Urethritis.
- Arthritis.
- Conjunctivitis.
- Mucocutaneous lesions.

Etiology

- *Infectious agents*—it may be due to pleuropneumonia like organism. Variety of infectious agents like *bedsonia*, *mycoplasma*, *chlamydia*, virus, etc.
- *Staphylococci*—it can be associated with staphylococci and in that case, it is called as *staphylococcal scalded skin syndrome*.

Clinical Features

- *Age and sex*—it is totally confined to men, usually between the ages of 20 to 30 years.
- *Symptoms*—the disease begins abruptly with diffuse erythema and fever.
- *Signs*—large flaccid bullae are formed which contain a clear yellowish fluid. The bullae rupture very easily leaving large areas of skin devoid of superficial epidermis.
- *Genital findings*—the urethral discharge is usually associated with an itching and burning sensation.
- *Arthritis*—arthritis is often bilateral, symmetrical and usually polyarticular.
- *Conjunctivitis*—conjunctivitis is often so mild as to be overlooked.
- *Skin lesion*—the skin lesions consist of red or yellow keratotic macules which eventually desquamate.

Oral Manifestations

- *Sites*—it is seen on the buccal mucosa, lips and gingiva.
- *Appearance*—the lesions appear as painless, red, slightly elevated areas, sometimes granular or vesicular with a white circinate border.

- *Palatal lesions*—the palatal lesions appear as small, bright red, purpuric spots which darken and coalesce.
- *Tongue lesions*—lesions on the tongue closely resemble geographic tongue. They may be mistaken as recurrent aphthous ulcers.

Diagnosis

- *Clinical diagnosis*—the clinical finding of peripheral arthritis which last longer than 1 month in association with urethritis, cervicitis will diagnose this syndrome.
- *Laboratory diagnosis*—there is leukocytosis, and elevated ESR. Biopsy of lesion shows parakeratosis, acanthosis, and polymorphonuclear leukocyte infiltration of epithelium, sometimes with microabscess formation.

Differential Diagnosis

- *Geographic tongue and stomatitis*—no skin changes, no visceral lesions are seen.
- *Pustular psoriasis*—Auspitz's sign present.
- *Behcet's syndrome*—no urethritis, aphthae with red halo.
- *Stevens-Johnson syndrome*—acute appearance, more severe clinical course, no arthritis or urethritis.
- *Benign mucosal pemphigoid*—blister formation, no urethritis, found in older patients.

Management

- *Spontaneous remission*—many patients undergo spontaneous remission.
- *Antibiotics*—in case symptomatic patient, doxycycline or minocycline may be given.
- *Analgesics*—nonsteroidal anti-inflammatory drugs are given to manage arthritis.
- *Immunosuppressive agents*—immunosuppressive agents like azathioprine and methotrexate are given in cases of most resistant cases.

Miscellaneous Oral Ulcers

Oral Ulcers Secondary to Cancer Chemotherapy

It may be due to direct effect on replication and growth of oral epithelium by interfering with nucleic acid and protein synthesis and leading to thinning and ulceration of oral mucosa, e.g. by methotrexate, or it may act indirectly by depression of bone marrow and immune response leading to bacterial, viral and fungal infections.

Clinical Features

- *Hair*—alopecia due to arrest of mitosis of the rapidly germinating hair root.
- *Stomatitis*—stomatitis occurs with diffuse inflammatory changes developed in mucosa.

- **Blebs**—distinct blebs or whitish areas result from decreased cellular division and retention of squamous cells.
- **Symptoms**—there is also burning sensation in the oral cavity with mucosal erosion.
- **Appearance**—in subsequent weeks, surface layer is lost and thin erythematous mucosa is present. Focal areas ulcerate and then become covered with tan yellow fibrous exudates (Fig. 17-37).



Fig. 17-37: Ulceration seen in palate after patient is given chemotherapy for the malignancy,

Management

- **Topical anesthetic**—topical anesthetic such as elixir of diphenhydramine or lidocaine combined with milk of magnesia.
- **Tetracycline suspension**—oral suspension of tetracycline which is used as mouthwashes and then swallowed.

Diffuse Gangrenous Stomatitis

It occurs in extremely debilitated patients, advanced diabetes, uremia, leukemia, blood dyscrasias, malnutrition state and heavy metal poisoning. It is sensitive painful oral lesions and very unpleasant odor. They are multiple and surrounded by thin inflamed margins. It is covered by dirty gray to yellow pseudomembrane that can readily be removed. It may be elliptical, linear and angular. There is tender to painful cervical lymphadenopathy. It is managed by systemic penicillin and hydrogen peroxide rinse.

Differential Diagnosis of Oral Ulcers

Short-Term Ulcers (Disappear within 3 weeks)

- **Ulcers secondary to systemic diseases**—patient history.
- **Ulcers from odontogenic infections**—identify the affecting tooth. Presence of sinus, gutta percha digital pressure in involved tooth or alveolus causes drops of pus to be expressed from the opening in ulcers.
- **Traumatic ulcer.**
 - Recurrent aphthous ulcers and recurrent herpes infection.

Persistent Ulcers (Longer than 3 weeks)

- **Metastatic tumor**—occurs in lower half of oral cavity, primary tumor elsewhere.
- **Low grade mucoepidermoid carcinoma**—not so common, occurs in posterolateral region of hard palate.
- **Gumma**—Mostly in midline of palate, rubbery consistency and serological finding.
- **Chancre**—it is reddish brown with copperhead colored halo.
- **Necrotizing sialometaplasia**—uncommon benign ulcerative inflammatory process of minor salivary glands found in posterior hard palate.
- **Keratoacanthoma**—seen in lower lip, rapid growth.
- **Major aphthous**—severe pain, inflammatory borders.

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Introduction

Pain and fear of pain continues to be the strongest motivation for people to seek dental care. Pain is a personal experience of the sufferer that cannot be shared and it wholly belongs to the sufferer. The head and face are subjected to chronic persistent or recurring pains more often than any other portion of the body. The dentist's responsibility in managing orofacial pain is twofold, first is the diagnosis, and second is the therapy.

The most frequent source of orofacial pain is dental disease and it has been estimated that toothache is a major health problem. Many conditions manifest as only orofacial pain with no other associated signs or symptoms. It has been clear that successful diagnosis of orofacial pain depend on (Fig. 18-1).

- *History*—an accurate and detailed history of the pain.
- *Clinical examination*—a detailed clinical examination of the face and associated organs.
- *Knowledge*—thorough knowledge of those conditions which may produce orofacial pain.

Pain is derived from word 'poine' meaning payment or penalty, which certain segments of our religious heritage have translated as being synonymous with punishment.



Fig. 18-1: Diagnosis of pain should be done with the help of three factors.

Severity of pain is a measure of the suffering it induces than the actual perception of the noxious stimulation. Low intensity pain may arise from very serious causes, whereas maximum intensity pain such as *tic douloureux* may arise from imperceptible causes.

Definition

- By IASP (*international association for study of pain*)—it is an unpleasant sensory and emotional experience associated with actual or potential tissue damage or described in term of such damage.
- It is an unpleasant emotional experience initiated by noxious stimuli and transmitted over a specialized neural network to the central nervous system, where it is interpreted as such.
- It is more or less a sensation of discomfort, distress or agony resulting from the stimulation of specialized nerve endings.
- It is a distressing sensation elicited by noxious stimuli of sufficient intensity acting on nerve endings.

Types and Nature of Pain

Pain may be judged subjectively or objectively. Subjective evaluation depends on the patient's description of the complaint. Objective evaluation relates to what the pain does to the patient physically as seen by change in vital signs, skin color, pupil size, and muscle effects.

There is definite relationship between intensity and duration of pain. The higher the intensity of the pain, the shorter the period of time that can be tolerated by the sufferer. Accordingly, low intensity pain can be sustained for upto several hours whereas maximum intensity pain can be tolerated just for few seconds. The higher the intensity of the pain, more likely it is to be intermittent.

Types According to Pain Intensity

- *Mild pain*—can be controlled by the use of simple analgesics.
- *Moderate pain*—can be controlled with narcotic analgesics.
- *Severe pain*—cannot be controlled by analgesics, but requires either elimination of the cause or interruption of the pain pathways.

According to Temporal Relationship and Duration

- *Intermittent*—pain of short durations and separated by wholly pain-free period.
- *Continuous*—pain of longer duration.
- *Protracted*—a painful episode that lasts for several days is usually described as protracted.
- *Intractable*—pain that does not respond to therapy.
- *Recurrent*—two or more similar episodes of pain.
- *Remission*—the pain-free interval between recurring episodes is called as remission.
- *Periodic*—pain that is characterized by regularly recurring episode is said to be periodic.

According to Qualities of Pain

- *Steady pain*—it flows as an unpleasant sensation.
- *Paroxysmal pain*—sudden attack or outburst of pain.
- *Bright pain*—stimulating quality.
- *Dull pain*—it has got depressing quality.
- *Itching*—it is a sub-threshold pain and usually is not described as pain at all. It may have a warm or even burning quality and can become intractable.
- *Pricking*—it has a sharp intermittent character of short duration like a pin pricking the skin.
- *Stinging*—it is more continuous and of higher intensity and quality.
- *Burning*—it gives a feeling of warmth or heat, when short and intense, it may have electric shock like feeling.
- *Throbbing*—pulsatile pain is timed to cardiac systole.
- *Aching*—it is the descriptive term most frequently used unless the pain is overshadowed by one of the other characteristic sensations.

According to Onset

- *Spontaneous*—if the pain occurs without being provoked.
- *Induced*—when some provocation causes the painful sensation.
- *Triggered*—when evoked response is out of proportion to the stimulus.

According to Pain Localization

- *Localized*—if the patient is able to clearly and precisely define the pain to an exact anatomical location.
- *Diffuse*—it is less well defined and somewhat vague and variable anatomically.
- *Radiating*—rapidly changing pain
- *Lancinating*—a momentary cutting exacerbation
- *Spreading*—gradually changing pain
- *Enlarging*—if pain progressively involves adjacent anatomical areas, it is called as enlarging.
- *Migrating*—if it changes from one location to another, the pain is described as migrating.

Neurophysiology of Pain

- *Structure of nerve*—nerve is a cord-like structure that conveys nervous impulse. It consists of a connective tissue sheath called as *epineurium*, which enclose bundles of nerve fibers, each bundle being surrounded by its own connective tissue sheath called as *perineurium*. Within each bundle the nerve fibers are separated by interstitial connective tissue called as *endoneurium* (Fig. 18-2).

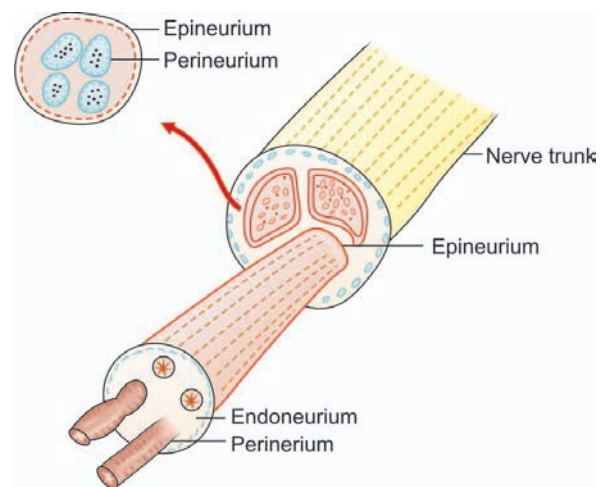


Fig. 18-2: Structure of nerve showing endoneurium and perineurium.

- *Structure of nerve fiber (Fig. 18-3)*—an individual nerve fiber consists of a central bundle of neurofibrils in a matrix of nerve protoplasm, called *axoplasm*, enclosed in a thin nerve tissue plasma membrane called *axolemma*. Each nerve fiber is covered by cellular nerve tissue sheath called as *neurolemma*. Some of these fibers also have a layer of fatty nerve tissue called the *myelin sheath*. Fibers with myelin sheath form the white nerve and those without the myelin sheath form the gray nerve.

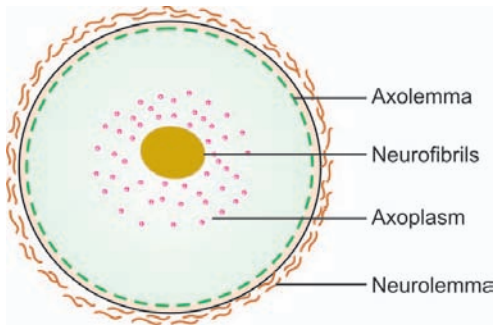


Fig. 18-3: Structure of nerve fiber diagrammatic representation.

- **Nodes of Ranvier**—constrictions called as *nodes of Ranvier* (**Fig. 18-4**) occur in myelinated nerves at intervals of about 1 mm. These nodes are caused by the absence of myelin material so that only neurolemma covers the nerve fiber. Those situated in the white substance are myelinated, whereas those in the gray substance are non-myelinated.

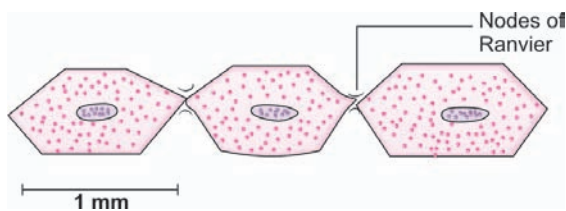


Fig. 18-4: Nodes of Ranvier.

- **Neuron**—the structural unit of the nervous system is the nerve cell or *neuron*. It is composed of a mass of protoplasm called nerve cell body (*perikaryon*), which contains a spherical nucleus (*karyon*) and gives off one or more processes.
- **Dendrites and axons**—protoplasmic processes from the nerve cell body are called as *dendrites* and *axons*. Dendrite (from the Greek word *dendron*, meaning tree) is a branched process that conducts an impulse towards the cell body (**Fig. 18-5**). An axon or axis cylinder is the central core that forms the essential conductor in a nerve cell and is an extension of cytoplasm from nerve cells.
- **Synaptic junction**—nervous impulses are transmitted from one neuron to another only at a synapse or a synaptic junction, where the processes of two neurons are in close proximity. All the afferent synapses are located within the gray substance of the CNS. Most of the primary synapse of nerve fibers, which carry pain, are located in the substantia gelatinosa of the cord and brainstem. The synapses that occur outside the CNS are those of efferent pre- and post-ganglionic autonomic fibers and these are located in the sympathetic ganglion.
- **Reflex arc**—some neural circuits are simple, i.e. an impulse from a sensory receptor being conveyed by the primary

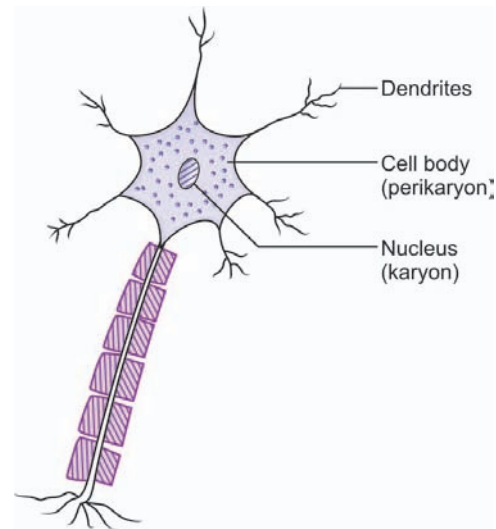


Fig. 18-5: Dendritic of nerve cell body.

afferent neuron to a synapse in the CNS with a secondary internuncinal neuron, which in turn synapses with a third efferent motor neuron, which will conduct the impulse peripherally to an effector organ such as a muscle. Such circuit formed by a chain of neurons in which stimulation is followed by an immediate and automatic response is called as *reflex arc*. The synapsing of a neuron with several other neurons is known as *convergence*. At the synapse, there may be cumulative effect, called *summation*. Intensification of response is known as *facilitation* and suppression of response is called as *inhibition*.

Types of Nerve Cells

Depending on the Number of Axons

- Unipolar
- Bipolar
- Multipolar

Depending on their Location and Function

- **Afferent neuron**—it conducts impulse toward the CNS.
- **Efferent neuron**—it conducts nervous impulse peripherally (**Fig. 18-6**).
- **Internuncinal neurons or interneuron**—it lies wholly within the CNS (**Fig. 18-7**).
- **Sensory or receptor neuron**—they are afferent in type, receive and convey impulses from receptor organs.
- **Motor or effector neurons**—they are efferent and convey nervous impulses to produce muscular or secretory effects.

- *Preganglionic neuron*—it is an autonomic efferent neuron whose nerve cell body is located in the CNS and terminates in an autonomic ganglion.
- *Postganglionic neuron*—it has its nerve cell body in the ganglion and terminated peripherally.

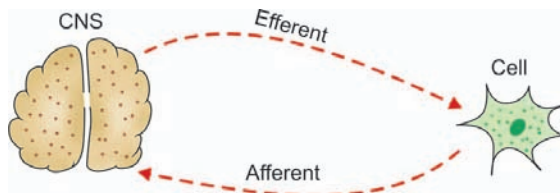


Fig. 18-6: Afferent and efferent nerve.

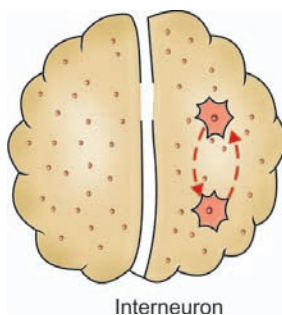


Fig. 18-7: Interneuron present in the brain.

Sensory Receptors

Sensory nerve fibers that respond to physical stimuli and convert them into nervous impulses for conduction towards the CNS are called sensory receptors.

There are three types:

Exteroceptors

They are stimulated by external environment and are thus located so as to become exposed to the external environment. Most impulses arising from these are sensed at conscious level. For example—

- *Merkel's corpuscles*—tactile receptors in submucosa of tongue and oral mucosa.
- *Meissner's corpuscles*—tactile receptors of skin.
- *Ruffini's corpuscles*—pressure and warmth receptors.
- *Krause's corpuscles*—cold receptors.
- *Free nerve endings*—perceive superficial pain and tactile sensations.

Interoceptors

They are located in and transmit impulses from the cavities of the body. Most impulses arising from these receptors are involved in involuntary functions of the body and are below conscious levels. For example—

- *Pacinian corpuscles*—concerned with perception of pressure.
- *Free nerve endings*—perceive visceral pain and other sensations.

Proprioceptors

They give information concerning the presence, position and movement of the body. For examples—

- *Muscle spindles*—mechanoreceptors found between the skeletal muscle fibers which respond to passive stretch of the muscle.
- *Golgi tendon organs*—mechanoreceptors in tendons of muscle.
- *Pacinian corpuscles*—receptors concerned with the perception of pressure.
- *Pressoreceptors*—receptors in periodontal ligament.
- *Free nerve endings*—perceive deep somatic pain and other sensations.

Pain Conduction

The conduction of an impulse by a nerve depends on the electrical potential that exists across the nerve membrane. The transmission of impulse is brought about by the flow of current across the membrane during the transition of the nerve from the resting to the active state (Fig. 18-8).



Fig. 18-8: Pain conduction showing resting stage, depolarization and repolarization.

Resting state

- *Ion concentration*—when the nerve is at rest, greater amount of anions (-ve) are present inside the cell membrane, whereas an equal number of cations (+ve) are gathered outside the membrane. The positively charged potassium ions are concentrated inside, while sodium and chloride ions are concentrated outside the membrane.
- *Creation of potential electrical difference*—the difference in respective ion concentrations across the nerve membrane

creates a potential electrical difference between the inside (negative) and outside (positive).

- *Resting potential*—thus, an unstimulated nerve at rest will have a resting potential, during which the membrane is said to be polarized with the inside being electrically negative relative to the outside. The resting potential of the nerve is maintained by the relative permeability of the cell membrane to potassium and its relative impermeability to sodium ions. Positively charged potassium ions are retained by the electrostatic attraction of the negatively charged nerve membrane. Chloride remains outside the nerve membrane because of the opposing electrostatic influence, forcing outward migration.
- *Sodium pump*—the maintenance of the resting potential is mainly because of an active mechanism called as 'sodium pump', which moves sodium from the area of lesser concentration inside the nerve to that of greater concentration outside. Because of greater concentration outside and the lesser concentration inside the nerve, the sodium ions tend to diffuse back across the membrane into the nerve while being pumped out. This pumping action controls the concentration of sodium on both sides of the membrane and thus maintains it in a polarized state. Thus, both concentration and the electrostatic gradients for sodium favor its inward movement, but, relative impermeability of the nerve during the resting state prevents the massive influx of this ion.

Depolarization

- *Activation of membrane by stimulus*—when a stimulus of sufficient intensity is applied to the nerve, the membrane is activated by the increased permeability of sodium, which diffuses through the membrane into the nerve cell.
- *Sodium diffusion*—the marked increase in the diffusion of sodium into the cell is followed by the passage of potassium out of the cell. This action is said to abolish the resting potential and depolarize the membrane.
- *Liberation of acetylcholine*—as the nerve is stimulated; there is a rapid passage of sodium into cell and a slower passage of potassium out of it. The alteration in permeability of cell membrane after the stimulus is a result of liberation of transmitter substance, acetylcholine, at the site of stimulation.

Repolarization

- *Change in permeability*—following depolarization, the permeability of the nerve membrane to sodium is again decreased, while the high permeability of potassium is restored.

- *Restoration of electrochemical equilibrium*—potassium moves freely out of the cell, thereby restoring the original electrochemical equilibrium and resting potential. Movement of both sodium ions into the cell during depolarization and potassium ions out of the cell during repolarization are both passive actions since these ions move along the concentration gradient.
- *Return of resting potential*—the return of resting potential occurs within 3 to 4 seconds after the initial stimulation.

Theories of Pain

Specificity Theory

In 1644, Descartes postulated this theory that pain system is a straight channel from the skin to brain. It was thought that this specific pain system carried message from the receptors to pain center in the brain.

Doctrine of Specific Nerve Energies

In 1842, Muller postulated this theory. This theory states that the brain receives information about external objects only from sensory nerves.

Pattern Theory

It was proposed by Goldscheiders. He emphasized that stimulus intensity and central summation are the critical determinants of pain. He proposed that the large cutaneous nerve fibers comprise a specific touch system whereas the small fibers converge on the dorsal horn cells, summate their input and transmit a pattern to the pain receptors which perceives the pain.

Peripheral Pattern Theory

A recent variation of pattern theory has been proposed by Weddell (1955) and Sinclair (1955) and it is called as peripheral pattern theory. This theory holds that all cutaneous qualities which are produced by spatiotemporal patterns of the nerve impulse rather than by the modality specific transmission route. All fiber endings are alike and pain is produced by intense stimuli to non-specific receptors.

Central Summation Theory

Livingstone suggested a specific neural mechanism, in which, with intense pathologic stimulation of the body, sets up a reverberating circuit in spinal internuncial pools that can be triggered by non-noxious stimuli leading to abnormal volleys that can be interpreted centrally as pain.

Sensory Interaction Theory

It is stated by Noordenbos and it states, the large fiber-pain inhibitory and small fiber-pain contributory, concepts with the two systems being in balance with one another. A decrease in the ratio of large to small fiber activity results in central summation and an increase in pain.

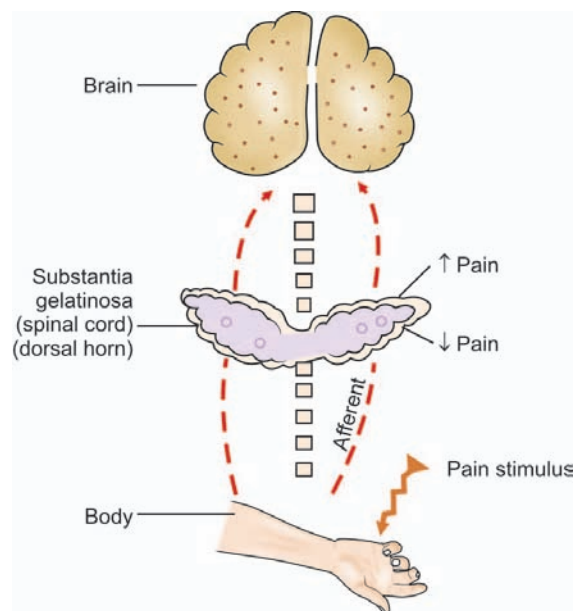
Gate Control Theory

This is the most accepted theory. It proposes that (Figs 18-9A and B):

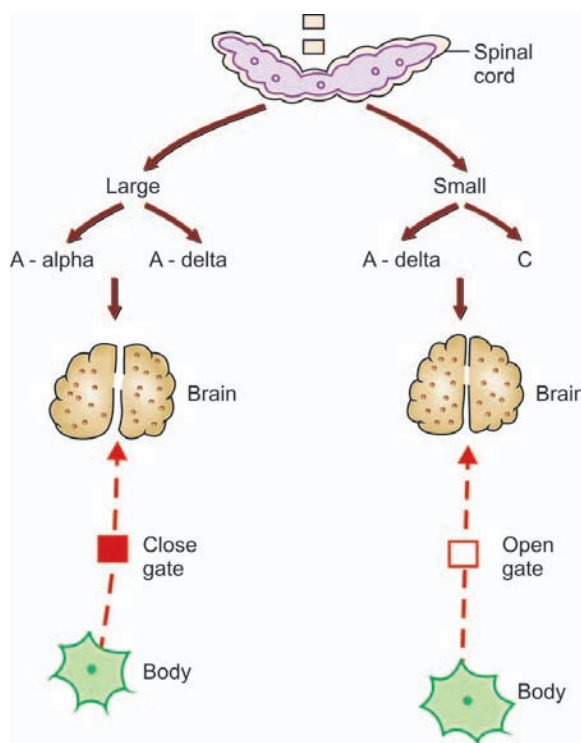
- *Neural mechanism in dorsal horns*—a neural mechanism in the dorsal horns of spinal cord (substantia gelatinosa) acts as a gate that can increase or decrease the flow of impulse from the periphery of the body to the brain before pain perception can be evoked. Afferent input is subject to the modulation of the gate before it can activate the central transmission cells and evoke pain perception as a response.
- *Alpha, delta and c fibers*—more active large (A-alpha and A-delta) fibers and the less active smaller (A-delta and C) fibers determine the degree to which the gate modulates sensory transmission. Activity of large fibers tend to inhibit transmission (close the gate) and small fiber activity tends to facilitate transmission (open the gate).
- *Impulse from brain*—it was further proposed that the control over transmission is affected not only by the gate mechanism but also by the impulse descending from the brain. Hence, the signal that triggers the system responsible for pain perception and patient response occurs when the T cell output reaches a certain critical level.

Measurement of Pain Intensity

- *Verbal communication*—this is communicated to you from the patient.
- *Visual analogue scale*—pain intensity can be measured by visual analogue scale. This scale consists of 10 cm lines. On this scale, there is marking from 0 to 10. 0 means no pain and 10 means very severe pain. Patient is asked to mark the line which represent pain.
- *McGill pain questionnaire*—this is useful in evaluation of pain. This questionnaire consists of 20 groups with 78 types of pain. In these groups, 1 to 10 is designed for assessment of sensory character, group 11 to 15 to assess affective character and from group 16 to 20 assess evaluative character of pain (Table 18-1).
- *Disability status*—this is very important in assessing the pain. Disability is lack of ability to function normally, physically and mentally.



A



B

Figs 18-9A and B: Gate control theory—a diagrammatic representation.

- *Multiaxial assessment of pain*—this includes 61 item questionnaires which measure adjustment to pain from cognitive behavioral pattern. This will help in getting profile of the patient. Patient profile can be dysfunctional (low activity with higher degree affective distress), distressed (patient think that other are supportive of their problems) and adaptive copers (patient with high level of social support).

Table 18-1: McGill pain questionnaire

Group 1	Burning	Vicious
Flickering	Scalding	Killing
Quivering	Searing	
Pulsing	Group 8	Group 15
Throbbing	Tingling	Wretched
Beating	Itchy	Blinding
Pounding	Smarting	
Group 2	Stinging	Group 16
Jumping	Group 9	Annoying
Flashing	Dull	Troublesome
Shooting	Sore	Miserable
Group 3	Hurting	Intense
Pricking	Aching	Unbearable
Boring	Heavy	
Drilling	Group 10	Group 17
Stabbing	Tender	Spreading
Lancinating	Taut	Radiating
Group 4	Rasping	Penetrating
Sharp	Splitting	Piercing
Cutting	Group 11	Group 18
Lacerating	Tiring	Tight
Group 5	Exhausting	Numb
Pinching	Group 12	Drawing
Pressing	Sickening	Squeezing
Gnawing	Suffocating	Tearing
Cramping	Group 13	Group 19
Crushing	Fearful	Cool
Group 6	Frightful	Cold
Tugging	Terrifying	Freezing
Pulling	Group 14	Group 20
Wrenching	Punishing	Nagging
Group 7	Grueling	Nauseating
Hot	Cruel	Agonizing
		Dreadful
		Torturing

- *Graded chronic pain severity scale*—this is given by Dworkin and LeReschae. It has got four grade of disability.
- *Quantitative sensory testing*—quantitative testing modalities include thermal, mechanical and electrical stimuli.

Classification of Orofacial Pain

- *Neuropathic pain*—orofacial pain associated with nerve dysfunction or abnormalities of the peripheral nerve. In this type pain is caused by the lesions affecting the intracranial but extra-pontine or extra-medullary portion of cranial nerves V, VII, IX, and X are classified as neuropathic
 - *Paroxysmal pain*
 - Trigeminal neuralgia
 - Geniculate neuralgia
 - Glossopharyngeal neuralgia
 - *Non-paroxysmal pain*
 - *Trauma*—pain occurring after trauma to nerve.
 - *Viral*—nerve getting involve due to virus.

- *Neoplasm*—neoplasm of the parotid gland, nasopharynx and acoustic neuroma may cause pain.
- *Non-neuropathic pain*—this type of pain involves nerves likely to be associated with the pain. It is subdivided into:
 - *Central origin*—this type of pain is caused by the lesions affecting the cord (central nervous system) is referred to as non-neuropathic pains of central origin.
 - *Neuronal damage*—pain results from neuronal damage in CNS.
 - *Phantom limb pain*—phantom limb pain and causalgia (a persistence burning sensation resulting from peripheral nerve trauma associated with various other phenomenon and curable by stellate ganglion block).
 - *Brain tumor and central lesion*—tumor, vascular lesion and other destructive lesions of the brain stem, pons, thalamus and cerebrum should be considered in the diagnosis of chronic orofacial pain.
 - *Other causes*—like thalamic syndrome of Dejerine and Roussy, tertiary neurosyphilis and multiple sclerosis.
- *Extra-neural origin*
 - *Dental*—this includes dental caries, exposed dentin and cementum, pulpal and periapical disease, periodontal disease, pericoronitis, impacted food, ANUG, faulty restorations, fractured teeth, traumatic occlusion, tooth eruption and exfoliation, unerupted and impacted teeth, retained root, dental cyst, post-surgical pain, post-injection pain, and other direct trauma to the teeth and aerodontalgia.
 - *Alveolar and adjacent tissue origin*—dry socket, sinusitis, various neoplastic and non-neoplastic, osteolytic lesions, Paget's disease, multiple myeloma etc.
 - *Musculoskeletal*—TMJ arthritis, TMJ myofascial dysfunction, myositis, muscle spasm and trismus, cervical muscle spasm, osteoarthritis, arthralgia of the cervical spine and tension headache.
 - *Vascular*—vascular headaches including migraine and cluster headache, toxic and metabolic vascular headache, hypertensive vascular changes, arterial diseases such as aneurysms, emboli, cranial arteritis (including giant cell arteritis, polymyalgia and immune arteritis), thrombophlebitis and carotidynia.
 - *Pain referred from outside the orofacial area*—otitis media, inflammatory or neoplastic diseases of the ear, diseases of major salivary glands (calculus, cyst and tumor), lesions of nasal and sinus mucosa, myocardial ischemia which may all cause pain in the orofacial area, often at some distance from the focus of disease.

Table 18-2: Second classification of orofacial pain

Superficial pain

- Cutaneous pain
- Mucogingival pain

Deep pain

- *Odontogenic pain*
 - Pulpal pain
 - Periodontal pain
- *Musculoskeletal pain*
 - *Muscle pain*
 - Local muscle soreness
 - Muscle splinting pain
 - Neoplastic myofacial pain
 - Muscle spasm pain
 - *Temporomandibular joint pain*
 - Disc attachment pain
 - Retrodiscal pad pain
 - Capsule pain
 - Arthrography pain
 - *Soft connective tissue pain*
 - Osseous pain
- *Vascular pain*
 - Atypical facial neuralgia
 - Cranial arteritis
- *Visceral pain*
 - True visceral pain
 - Reflex sympathetic dystrophy

Neurogenous*Traumatic neuroma pain*

- *Neuritis pain*
 - peripheral neuritis
 - herpes zoster
- *Neuralgia pain*
 - Idiopathic neuralgia
 - Symptomatic neuralgia

Psychogenic pain

- Psychogenic intensification pain
- Conversion hysteria pain
- Delusional pain

- *Pain of unknown nature*—pain that arises outside the peripheral nerve and only affects the nerves and their receptors secondarily (e.g. pain of dental caries, periodontal disease and mouth ulcers).

Acute Pain

Acute pain may be considered to be a protective mechanism for the body, by stimulating the sympathetic nervous system, is often accompanied by the automatic signs of stress and anxiety. It serves as a diagnostic value to the clinician in determining the nature and site of the disturbance.

Chronic Pain**What is chronic pain?**

It is persistent of pain after healing. Pain of more than 6 months duration may be considered as chronic pain.

Table 18-3: Third classification of orofacial pain

Typical neuralgias

- Trigeminal
- Glossopharyngeal
- Geniculate
- Superior laryngeal
- Post-herpetic

Atypical neuralgias

- Migraine
- Spheno-palatine neuralgia
- Migranous neuralgia
- Ciliary's neuralgia
- Petrosal neuralgia
- Carotodynia
- Cluster headache

Typical facial pain

- *Pain of extra-cranial origin*
- Dental disease
- Ocular disease
- Ear, nose and throat disease
- Salivary gland disease
- TMJ syndrome
- Musculoskeletal disorders of the head and neck
- Cranial arteritis
- Thyroid disease
- Angina pectoris
- *Pain of intra-cranial origin*
- Trigeminal neuropathy
- Tumor of middle cranial fossa and Gasserian ganglion
- Cerebellopontine angle tumor
- Thalamic central pain syndrome
- Trigeminal neuralgia in multiple sclerosis

Atypical facial pain

- It refers to mixed group of conditions diagnosed by exclusion of other typical patterns of facial pain. The conditions are characterized by poorly localized, deep, constant discomfort which is non-anatomic, non-dermatomal and is frequently described by the patient in vague terms which gives psychogenic basis for the disorder.

Table 18-4: Fourth classification of orofacial pain

Acute

- *Dental*—pulpitis, cracked tooth syndrome
- Gingivitis, periodontitis, pericoronitis
- *Mucosal*—various causes of ulceration
- *TMJ*—traumatic acute dysfunction
- *Maxillary sinus*—sinusitis and carcinomas
- *Salivary gland*—acute sialadenitis
- *Ear*—otitis externa and media
- *Tonsils*—quinsy and carcinoma
- *Referred*—cardiac, angina, cervical spondylitis

Chronic

- *Neuropathic*—trigeminal and post-herpetic neuralgia
- *Nociceptive*—cancer, osteoarthritis, rheumatoid arthritis
- *Chronic and recurrent idiopathic orofacial pain*—TMJ dysfunction syndrome, atypical facial pain, atypical odontalgia, oral dysesthesia, tension headache, migraine, facial migranous neuralgia.

Chronic pain does not serve any apparent biological function and is psychologically destructive. It is not self-limiting and appears to be permanent. Nowadays, any persistent pain more than 3 months of duration is called as chronic pain. Merskey et al described chronic pain as 'persistent pain that is not amenable, as a rule to treatment based on specific remedies or to the routine methods of pain control such as non-narcotic analgesics'.

Pathophysiology

- *Inflammatory origin*—many orofacial pains of chronic are inflammatory in origin. The inflammatory response to tissue damage, results in the production of pain, edema, local increase in temperature, redness and loss of function.
- *Neuroplasticity*—neurons are capable of altering the structure and function in response to stimuli. This will result in new stimulus response relationship.
- *Reduction in stimulation threshold*—this will result in activation of neurons without noxious stimulus.
- *Alteration in temporal pattern*—due to this transient stimulus may lead to sustained burst of activity.
- *Greater response*—there is increase in general response of motor neurons. This will result in more response to noxious stimuli.
- *Expansion of receptive field*—this will result in wider area may get involve with the stimuli.

Clinical Manifestation

- *Hyperalgesia*—changes in CNS result in the clinical manifestation of hyperalgesia. Hyperalgesia is characterized by spontaneous pain, decreased pain threshold and an increased magnitude of perceived pain for any given stimulus.
- *Allodynia*—there is pain due to stimulus that does not normally cause pain.
- *Psychosocial disturbance*—patient suffers from the depression, anxiety, and social isolation.
- *Dysfunction*—due to chronic pain normal activity of patient is decreased.

Superficial Pain

Superficial pains have a bright, stimulating quality and can be correctly located by the patient so that he is aware of precisely where it hurts. He is able to describe the location of the pain with anatomical accuracy and thus the site of pain and site of origin are identical.

Superficial pain is primary hyperalgesia, i.e. the structure is painful owing to lowered threshold from local cause. The reaction is proportional to the intensity of the stimulus; it lasts as long as the stimulus and there is no reference to pain to other structures.

Types and Clinical Features

- *Cutaneous pain*—it is characterized by pricking or burning sensation.
 - *Pricking pain*—pricking pain will occur if the stimulus is slight and short duration.
 - *Stinging pain*—if the stimulation is slight but sustained or rapidly repeated a slight stinging sensation is felt.
 - *Burning pain*—severe stimulation can cause burning pain.
- *Mucogingival pain*—it differs from cutaneous pain as it does not show the typical pricking; burning pattern of cutaneous pain. The reason for this is that the number of thick fibers innervating the oral mucosa is negligible as compared to that of skin. The entire oral mucosa feels a stinging and burning sensation. Pain arising from the oral tissues can be precisely located by the patient. Mucogingival pain can be generalized, isolated, referred and neurogenous.
 - *Generalized mucogingival pain*—it is caused by following factors:
 - *Local causes*—irritating effect of substances like food, liquid, mouth washes, dentifrices, and medications. Generalized mucogingival pain may occur due to abrasive effect of excessive rubbing of the tongue and cheek. In this, various signs of hyperemia, inflammation can be seen at the site of pain.
 - *Local infection*—widespread local infection of mucogingival tissue can cause generalized superficial somatic pain. Signs of inflammation and ulceration make the cause of pain obvious.
 - *Glossodynia, burning mouth and burning tongue*—it may cause steady, continuous typical superficial somatic pain. The location of pain corresponds to the area of greatest movement. Secondary infection of the irritated tissue may complicate the symptoms. Xerostomia may also cause this condition.
 - *Massive superficial injury*—traumatic effect from mechanical, thermal or chemical cause such as scalding with hot foods or liquids, excessive smoking or use of tobacco, abrasion from tooth brushes, etc. may cause pain.
 - *Allergic response*—it includes stomatitis medicamentosa and stomatitis venenata. They may produce rather generalized mucogingival pain. These conditions are usually accompanied by the objective sign of tissue change and the cause can be known by careful history taking.
 - *Pregnancy*—pregnancy and perhaps the ingestion of oral contraceptive drugs cause oral manifestations which may cause pain.

- *Systemic disease*—systemic diseases may cause oral manifestations that affect the mucogingival tissues causing generalized superficial somatic pain, for example, nutritional deficiency, intoxication, anemia, other blood dyscrasias, pellagra, diabetes, and pemphigus.
- *Isolated mucogingival pain*—various isolated lesions, like herpes simplex, oral ulcerative lesion of aphthous stomatitis, ulcer form dental appliances, can cause isolated pain in the mucogingival tissue. Whenever the periosteal and osseous structures become involved, pain of mixed quality is generally expected.
- *Referred mucogingival pain*—according to central excitation theory, the input of deep pain impulses may stimulate other neurons that mediate pain causing referred pain. If that neuron mediates superficial pain impulse then referred pain will have characteristics of superficial somatic pain. Thus, deep pain may secondarily induce mucogingival pain. Referred pain in the mucogingival tissue may occur as a direct result of deep pain source such as toothache, earache, sinusitis, temporomandibular arthralgia and vascular pain.
- *Neurogenous mucogingival pain*—neurogenous pains present clinical characteristics simulating those of the superficial somatic pain. For example, painful neuroma, neuritic pain, neuralgia pain.

Deep Pain

Sensory innervations of the deeper structures of the body supply the cortex with constant inflow of information regarding all internal functioning of the body. Deep pain has a dull, depressing quality. Deep pain is less accurately localized by the patient and the patient's anatomical description of where he feels the pain may be quite diffuse. Deep pain may not be proportional to the stimulus. Some deep pains respond rather faithfully to stimulus but not as faithfully as superficial pain. One of the clinical characteristic of the deep pain is central hyperexcitability. Deep pain input tends to cause referred pain, secondary hyperalgesia, localized autonomic effects and secondary myospasm activity.

Pains of Dental Origin

Dental pain is usually described as an aching sensation; sometimes throbbing, and when severe, may have a burning quality.

- *Pulpal pain*—pulpal pain may be classified as acute, chronic, and recurrent or mixed with periodontal element. Pulpal pain may resolve, or become chronic.
- *Acute pain*—the cause of acute pulpal pain is noxious stimulation of the pulpal receptors. This type of pain

is not completely localized by the patient. The cause for acute dental pain is deep caries, erosion, fracture or splitting tooth. This will help in identification of the offending tooth. As soon as pulp is exposed, it becomes painful in contact with saliva or air. The pain threshold of all deep receptors and nerve fibers that mediate pain is lowered by sustained hyperemia or inflammation. Thus, dental pulp that is hyperemic or inflamed is hypersensitive to all stimuli including electric stimulation, thermal shock, probing and percussion. As the inflammatory process progresses spontaneous toothache may occur without any outside provocation. Acute pulpal pain may range from occasional hypersensitivity caused by sweet and other minor stimulants to spontaneous violent throbbing toothache of intolerable intensity. It may be induced by any type of irritants or be wholly spontaneous. It may be increased by both heat and cold or increased by heat and relieved by cold. Pain frequently occurs at night when the patient lies down or when the patient leans down, most probably because of an increase in blood pressure. The difference between a hyperemic pain caused by thermal reaction and acute pulpitis is that in acute pulpitis the pain does not necessarily resolve when the irritants are removed but continue to ache for few minutes and possibly for hours. The usual relief for advanced acute pulpitis is cold water.

- *Chronic pain*—injured pulpal tissue may progress from an acute to chronic inflammatory phase. It results from a continued low grade infection. When chronic pulpitis occurs, pain response changes from extremely variable character of acute pulpal pain to milder and less variable vague discomfort that may not be described as pain at all. Tooth may become symptomless.
- *Toothache from non-dental cause*—pain referred to teeth from inflamed nasal mucosa, nonspastic myofacial pain especially involving temporalis and masseter muscle, myofacial pain-dysfunction syndrome, pain of the heart muscles as a symptom of cardiac disease. Neuritic pain such as those involving the maxillary teeth from inflammatory involvement of the superior dental plexus by the disease of the antrum and from neuritic conditions of the inferior alveolar nerve.
- *Periodontal pain*—the receptors of the periodontal ligament are capable of rather precise localization of the stimulus, therefore periodontal pain of all types present no difficulty in diagnosis. The offending tooth is identified by applying the pressure to it laterally or axially. It may occur as a primary periodontal inflammatory condition due to local causes such as trauma, occlusal stress, and contact with an adjacent

embedded tooth, dental treatment like dental prophylaxis, occlusal interference, over contoured or under contoured, proximal contact area. It may result from spread of pulpal infection or by direct extension from a nearby inflammatory condition involving an adjacent tooth, the maxillary antrum, or a spreading osseous infection. When periodontal pain involves several teeth, especially opposing teeth, then occlusal oversteering either by clenching or bruxism should be considered. Oversteering of posterior teeth may occur as a result of decreased vertical height of the mandibular ramus due to osseous fracture or degenerative changes in the TMJ.

- *Cracked tooth syndrome*—patient complains of pain ranging from mild to excruciating, at the initiation or release of the biting pressure. It can mimic the condition as severe as trigeminal neuralgia. A crack may involve enamel and dentin only or it may also involve the pulp and symptoms will vary accordingly. Pain occurs due to fluid movement within the dentinal tubules causing stimulation of sub-odontoblastic nerve fibers. The fluid movements are induced by pressure changes when biting with the offending cusp. If the crack involves dental pulp, direct bacterial invasion will occur with predictable pulpal inflammation and resultant pulpitis pain. Close examination of the crown of the tooth may disclose a crack in enamel, which may be better visualized by using a dye or by trans-illuminating the tooth with fiber-optic light. Crack can be confirmed by selective biting pressure using a cotton roll or a small wooden stick to allow selective localization of such pressure. It is treated by splinting of the offending cusp with a cusp protecting restoration or by removing the split cusp and then restoring the tooth.
- *Aerodontalgia*—it occurs due to effect of change in the altitude. Persons experience pain during high altitude flight or during deep sea diving. At ground level, the tooth is asymptomatic. It occurs due to subclinical pulpitis. It may occur in patients with improper obturation as improperly obturated root canal may expand during flight or during diving which creates pressure in the periapical nerve bundles and produce pain.

Musculoskeletal Pain

Pain arising from musculoskeletal structures present clinical characteristics by which they may be differentiated from other causes of deep pain; the pain relates reasonably and logically to use, movement and the demand of function. The pain can be provoked or aggravated by manipulation of the structure involved. The pain is usually but not always accompanied by some dysfunction.

Muscle pain frequently occurs in otherwise normal area, as a result of reflex protective mechanism and myofacial triggers. Muscle pain is usually felt as a non-pulsatile variable aching sensation, sometimes having a boring quality. The pain may become more lancinating and may occur spontaneously and in response to stretching, contraction, manipulation or manual palpation. Sometimes, pain is not more than a feeling of pressure whereas sometimes its intensity is increased. It may be transitory, persistent, constant, intermittent or recurrent.

Dysfunction may be expressed as tightness and weakness or impairment of muscle function such as stiffness, rigidity, swelling, and tenderness on palpation. The fact that movement and functioning modify the pain and stiffness are clinical indications of the presence of muscle pain.

- *Mechanism of muscle pain*—the mechanism is related to accumulation of metabolites after excessive use of muscles. This will result in distortion of blood vessels within the muscle, causing ischemia and hyperemia. Forceful and sustained contraction causes vasoconstriction of the relevant nutrient arteries resulting in pain.
- *Causes*—various factors like unusual yawning, biting, chewing, and strained sleeping position will cause muscle pain. Other factors which can cause muscle pain are minor blows, playing a musical instrument, period of sustained emotional tension, bruxism, excessive or prolonged opening of mouth for dental treatment, and use of local anesthetics.
- *Local muscle soreness*—it is a local condition with a local cause. It is a primary hyperalgesia with lowered pain threshold due to local factors such as strain, injury, abusive use, infection, and inflammation. When the dysfunction involves elevator masticatory muscle, trismus may result, restricting the normal mouth opening. When the muscle becomes inflamed as a result of injury or infection, the pain may relate to irritation and pressure of inflammatory exudates.
- *Muscle splinting pain*—it is defined as rigidity of the muscle occurring as a means of avoiding pain caused by movement of the part. It is a reflex protective mechanism whereby skeletal muscle becomes hypertonic and painful when contracted. It differs from muscle spasm in which contraction is sustained even when the muscle is at rest. There is pain and restriction of movement during active contraction of the involved muscle with little or no evidence of pain and restriction when purely passive movement is executed. Splinting of masticatory muscle may occur as a protective mechanism in conditions such as toothache, occlusal interference, sensitive overstressed teeth, and effect of local anesthetics, surgery, and trauma. Sustained splinting may develop into muscle spasm.

- *Nonspastic myofacial pains*—there is no muscle spasm and pain is the only complaint. Certain muscles tend to develop trigger areas within the muscles or tendons. When these sites are stimulated by ordinary function (contraction and stretching) pain impulses are generated and pain may or may not be sensed in the muscle. There is secondary referred pain which is felt in structures located at some distance from the trigger site and thus, the pain may be felt in adjacent normal structures. It may be caused by atrophied muscles due to inactivity, sustained emotional tension, illness, nutritional deficiency. Once the myofacial triggers develop within the muscle, they tend to persist as a source of intermittent and recurrent pain when the triggers are stimulated.
- *Zones of pain reference from myofacial triggers*
 - *Masseter muscle*—the masseter muscle refers to the ear, TMJ and mandibular teeth.
 - *Temporalis muscle*—the temporalis to the temple, orbit and maxillary teeth.
 - *Medial pterygoid*—the medial pterygoid to the infra-auricular and post mandibular area.
 - *Lateral pterygoid*—the lateral pterygoid to the TMJ.
 - *Sternomastoid*—the sternomastoid to the ear, pre-auricular area and widely throughout the face.
 - *Splenius capitis*—the splenius capitis to the parietal region.
 - *Trapezius*—trapezius to the neck, temple and frontal region.

Temporomandibular Joint Pain

Pain that results from TMJ and capsular ligament of TMJ is identified as masticatory pain. The pain is considerably more stable than myalgia. It can occur as intermittent arthralgia (occurs due to stress, pressure or movement) or continuous arthralgia (insidious, changes slowly, persistent and resists therapy). Central hyper-excitability tends to occur causing referred pain, secondary hyperalgesia and myospasm. The pressure bearing surfaces of the joint are non-painful due to absence of sensory receptors and nerve fibers. Pain can occur in disc when impinged upon and displaced sufficiently to stimulate receptors in the ligamentous attachment of the disc to the medial and lateral poles of the condyle.

- *Retrodiscal pad pain*—inflammation of the tissue that comprises the retrodiscal pad is called as retrodiscitis. If the condyle is displaced posteriorly because of posterior overclosure from missing teeth or from trauma, the resultant inflammatory swelling will displace the condyle anteriorly causing acute malocclusion described as contralateral premature contact of the anterior teeth with disocclusion of the posteriors ipsilaterally.

- *Capsular pain*—inflammation of the fibrous and synovial capsule is termed 'capsulitis'. Being inflammatory, pain results whenever the capsule is stretched by translatory movement of the condyle.

Vascular Pain

Pain of this type occurs because of noxious stimulation of the afferent nerve fibers that supplies sensation to blood vessels. The characteristics of vascular pain can be differentiated from other types of deep somatic and visceral pain. They are:

- *Spontaneous pain*—the pain occurs spontaneously without regard from the demand for function. Exacerbations occur spontaneously and without any provocation.
- *No pain on provocation*—provocation of the site of pain does not initiate or aggravate the pain.
- *Recurrent and periodic behavior*—the behavior of pain is typically recurrent and periodic.
- *No dysfunction*—no dysfunction accompanies the pain other than the inhibitory influence of pain and the effect of secondary muscle spasm as a central excitatory effect.

Wolf has indicated that there are several conditions that cause discomfort in vascular pain syndrome. These are dilatation of blood vessels, local edema at the painful site, edema of the vessel walls and perivascular tissues, and associated muscle pain especially in the occipital area.

The distinctive feature of vascular pain is its primary pulsatile or throbbing quality. Greater the amplitude of vascular dilatation, more pronounced the throbbing quality of the pain. This quality may be very slight at times and may be masked by muscle pain but is usually present during exacerbation. Although not invariably, there is an emotional factor that accompanies vascular pain syndrome. Frustrations, fatigue, feeling of insecurity with tension in the individual appears to set the stage for a painful episode.

Migraine

It is also called as *migraine syndrome* or *migraine headache*.

Etiology

- *Trigeminovascular neuron activation*—there is activation of trigeminovascular neurons surrounding a cephalic blood vessel due to vasoconstriction. This will lead to cerebral ischemia followed by compensatory vasodilation with subsequent pain and cerebral edema.
- *Hereditary*—it has got autosomal dominant inheritance pattern.
- *Triggering factors*—Diversity of factors is thought to be triggering the attack. It includes dietary factor (chocolate,

missing meal, aged cheese), psychogenic (stress, anxiety, depression), hormonal (menstruation, ovulation, oral contraceptive), sleep disturbance, physical (glare, flashing light, fluorescent light, odors high altitude), drugs (nitroglycerine, histamine, reserpine, hydrazine, ranitidine, estrogen) and miscellaneous (head trauma, fatigue, physical exertion).

Types

- *Migraine with aura*—in this, an aura or pre-headache period occurs. This is characterized by visual disturbances like scintillation (seeing sparks), scotoma (partial or complete loss of light perception). There is also aphasia (loss of ability to express thoughts), and numbness over one side of the face and arm.
- *Migraine without aura*—in this, no pre-headache period occurs.

Clinical Features

- *Age and sex distribution*—it begins at young age, and primarily affects women in a ratio of four to one.
- *Location*—headache is unilateral and felt in temporal, frontal, and orbital region. Rarely, it can be felt in parietal, postauricular and occipital areas.
- *Onset*—headaches often begin immediately or soon after awakening. The attack may be recurrent or episodic with variable frequency, usually 1 to 4 in a month.
- *Nature of pain*—it is started with mild headache which later increases in severity. It is of throbbing quality at the peak.
- *Associated symptoms*—there are other symptoms like nausea, vomiting, anorexia, sensitivity to light and sound, and mood changes.

Diagnosis

- *Clinical diagnosis*—pre-headache symptoms with presence of trigger zone with throbbing type of pain.

Management

- *Severe attacks*—these are controlled by ergotamine tartrate combined with caffeine, aspirin, acetaminophen, belladonna and Phenobarbital.
- *Mild cases*—treated by methergine, beta adrenergic agents and calcium channel blockers and serotonin receptor agonists.
- *Drug therapy*—drug which is used in treatment of cluster head are ergotamine (2 mg sublingual tablet), sumatriptan (50-100 mg tablet, agonist to 5-HT_{1D}) will give relief from the symptoms. Another drug which can be used are naratriptan (2.5 mg tablet), almotriptan (12.5 mg tablet), rizatriptan (5-10 mg tablet), zolmitriptan (2.5 mg tablet).

- *Non-pharmacological management*—non-pharmacological management includes diet control, stress management, sleep regulation and pressure on ipsilateral carotid artery.

Cluster Headache

It is also called as *migranous neuralgia*, *sphenopalatine neuralgia*, *histaminic cephalgia*, *periodic migraine* and *Horton syndrome*. It is an uncommon. It is the most severe headache and has been referred to as suicide headache. It is called as cluster as attacks occur in groups or cluster.

Etiology

- *Vascular cause*—this disease is caused by abnormal hypothalamic function, head trauma, and abnormal release from mast cells.
- *Triggering factors*—headache can be initiated by alcohol, cocaine, and nitroglycerine.

Clinical Features

- *Age and sex distribution*—more common in third to fourth decade of life with strong male predilection.
- *Location*—the deep intense pain may last for 15 minutes to 3 hours, which is unilateral and involves the peri-orbital area, often radiating to the ipsilateral temple and maxilla including the teeth. Pain is present in the distribution of ophthalmic nerve.
- *Nature of pain*—pain is paroxysmal, burning, and lancinating without trigger zone.
- *Alarm clock headache*—Attack develops regularly, usually once at day over prolonged periods and some patients at same time in day for this reasons, is referred to as 'alarm clock' headache.
- *Associated features*—associated symptoms, such as lacrimation from the eye, nasal congestion, rhinorrhea, forehead and facial sweating, miosis and eyelid edema, may be seen. Paresthetic sensation of skin over the lower half of the face has been also reported.

Diagnosis

- *Clinical diagnosis*—alarm clock headache without trigger zone will diagnose this condition.

Management

- *Drug therapy*—drug which is used in treatment of cluster head are ergotamine (2mg sublingual tablet), prednisone, lithium carbonate, indomethacin, methylsergide maleate, and verpamil, sumatriptan (50-100 mg tablet, agonist to 5-HT_{1D}) will give relief from the symptoms.

- *Oxygen inhalation*—inhaling oxygen may shortened attacks of symptoms.
- *Surgical*—surgical interventions have been tried. It includes trigeminal sensory rhizotomy, superficial petrosal neurectomy, gamma knife radiosurgery, and decompression of the nervus intermedius.

Temporal Arteritis

It is also called as cranial arteritis or giant cell arteritis. It most commonly affects the head and neck vessels.

Etiology

- *Inflammation and obstruction*—it is caused by inflammation and obstruction of the temporal and other cranial arteries.
- *Autoimmunity*—it may be caused by autoimmunity of elastic lamina of the artery.
- *Genetic*—there may be genetic predisposition for this disease.
- *Preceding factors*—extraction of infected tooth may cause exacerbation of pain.

Clinical Features

- *Age and sex distribution*—it occurs exclusively in individuals over the age of 50 years. It affects women more frequently than males.
- *Onset*—onset may involve one or both temporal areas.
- *Symptoms*—severe throbbing headache that often has abrupt onset and pain, usually being on one temple but occasionally involve the whole side of face. Headache may be associated with hyperesthesia. There is pain on mastication, pain in teeth, jaws and zygoma region due to involvement of internal and external maxillary artery. Patient may feel pain on chewing and talking also.
- *Nature of pain*—moderate to severe headache of deep, aching, occasional throbbing or burning quality may be present.
- *Radiating point*—pain present may radiate from the temporal region to the neck, maxilla, mandible or face.
- *Polymyalgia rheumatica*—sometimes, there is aching and stiffness of the muscle of the shoulders and hips which is often termed as ‘polymyalgia rheumatica’.
- *Tongue involvement*—pain present in tongue associated with blanching and even gangrene, which occurs because of involvement of lingual arteries.
- *Associated features*—there may be swollen and tender scalp arteries, usually the superficial temporal artery.
- *Complication*—if prompt treatment is not carried out, it may result in eye pain, photophobia, diplopia and blindness.

Diagnosis

- *Clinical diagnosis*—polymyalgia rheumatica with severe headache in temporal region.

Management

- *Corticosteroids*—corticosteroids is the treatment of choice and clinical manifestations subside within few days. Most commonly given is glucocorticoid therapy (75 gm daily). Steroids doses are reduced by 5 mg per week to maintenance dose 10 mg daily for 3 months.

Tension Headache

This is the most common type of headache with maximum number of adults suffers from it.

Etiopathogenesis

- *Psychophysiologic changes*—Psychophysiologic changes like stress, anxiety, and depression may cause this type of headache.
- *Masticatory muscle contraction abnormalities*—abnormalities of masticatory muscle contraction will also result in tension type headache.

Clinical Features

- *Age and sex distribution*—this is more commonly seen in adults and women are affected more commonly than male.
- *Nature of pain*—the pain is episodic, which is thought to be due to psychological or physical stressful events or of chronic type related to stressful daily life. It is usually slow budding pain. Pain is bilateral, dull, aching sensation, of mild to moderate intensity, with a pressing or tightening quality.
- *Physical activity*—unlike migraine, physical activity does not worsen the pain and nausea is absent.
- *Tender areas*—patients often have tender areas on the scalp and neck and palpation may reveal tender nodules in the cervical and trapezius muscle groups. These nodules may be trigger points and have the capacity to refer to the face and head area.

Diagnosis

- *Clinical diagnosis*—by looking at the symptoms, it is easy to make diagnosis of tension type headache.

Management

- *Psychological management*—simple counselling, hypnosis, relaxation and biofeedback measure are helpful.

- *Analgesic*—NSAID like aspirin are used commonly to treat tension type headache.
- *Tricyclic antidepressant*—this is also useful for tension type of headache. Most commonly given drugs are amitriptyline, and doxepin.

Chronic Paroxysmal Hemicranias

Chronic paroxysmal hemicrania is vascular type of headache which mimic migraine headache.

Clinical Features

- *Age and sex distribution*—it is more commonly seen in adulthood and more common in female.
- *Onset*—it has rapid onset characterized by unilateral head pain lasting for minute to hours.
- *Site of pain*—temporal and orbital region are site of pain. Jaw and face are also affected.
- *Associated symptoms*—there is also lacrimation, rhinorrhea, nasal stuffiness and swelling of painful area.

Diagnosis

- *Clinical diagnosis*—diagnosis is made from patient history, area of pain location and high frequency pain.

Differential Diagnosis

- *Cluster headache*—it is an attack with same characteristics of pain and associated symptoms and signs of a cluster headache, but they are shorter lasting, more frequently, occur in females, and there is absolute effectiveness of indomethacin.

Management

- *Indomethacin*—it is given in dose of 25 to 150 mg daily. It is effective in all cases of chronic paroxysmal hemicranias.

Carotidynia

A rare form of pain of vascular origin can arise from carotid artery and is therefore, known as carotidynia. It is characterized by a tender, throbbing, often swollen carotid artery on the affected side with radiation of pain to branches of the external carotid artery. The carotid tenderness is most apparent at the bifurcation or under the mandible.

Atypical Facial Pain

It refers to a mixed group of conditions which are defined and diagnosed by exclusion of other typical patterns of facial pain. It is also called as *typical facial neuralgia*, *idiopathic facial pain*, *atypical trigeminal neuralgia* and *trige-*

minal neuropathic pain. In this condition there is occurrence of strong emotional overtones of the conditions. It is usually psychogenic and occurs in patients who suffer from depressive reaction, hysteria, or schizophrenia (Fig. 18-10).

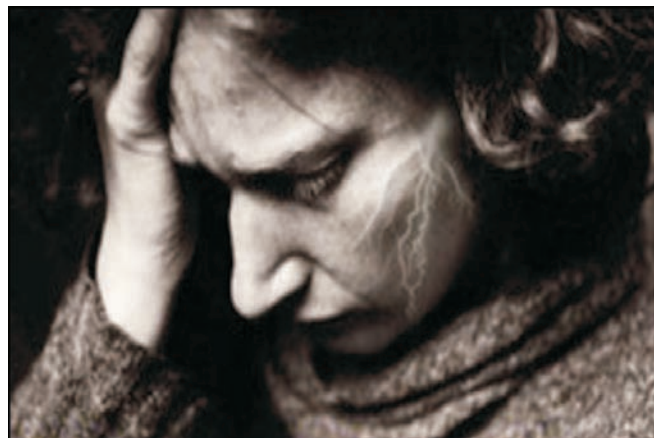


Fig. 18-10: Facial pain may be very distressing for the patient.

Clinical Features

- *Age and sex distribution*—it is more common in sixth decade with women more commonly affected.
- *Characteristic of pain*—the condition is characterized by pain that is deep, poorly localized, and vaguely described by the patient. Pain is often boring, pressing, pulling, burning, or aching. The distribution of pain is not anatomical. In general, pain is constant.
- *Referred pain*—pain is referring to temple, neck and occipital area.
- *Hot spot*—the mucosa of affected person may contain zone of increased temperature and bone marrow activity showing hot spot on technetium 99m MDP bone scan.

Diagnosis

- *Clinical diagnosis*—the diagnosis of 'atypical facial pain' can be justified where, there is an unusual character of pain, the pain does not have an anatomic distribution, the symptoms indicate some emotional illness or when significant organic development is lacking.
- *Disease to be ruled out for diagnosis of atypical facial pain*—sinus allergy, cracked tooth syndrome, headache with referred pain to face, bone impingement on nerve, Myofascial pain, neuralgias, temporomandibular joint disorders and trauma to nerve.

Management

- *Opioid analgesics*—these can be given in this patient. But their effectiveness may be diminished over period of time.
- *Tricyclic antidepressants*—tricyclic antidepressant like amitriptyline, nortriptyline is given in many cases. This

drug should be given cautiously with patient suffering from coronary heart disease.

- *Other therapy*—other therapy like psychotherapy, behavior modification, and transcutaneous electrical nerve stimulation and sympathetic nerve block are helpful in atypical facial pain.

Neurogenous Pain

Characteristics of Typical Neurogenous Pains

- *Bright quality*—the pain has a bright stimulating quality.
- *Exact location of site of pain*—the site of pain is accurately localized by the patient.
- *Analgesic blocking*—site of pain does not necessarily identify the site of origin (except traumatic neuroma). Site of pain bears a precise anatomical relationship to the site of origin, so that analgesic blocking is useful diagnostically.
- *Intense pain*—pain is extremely intense compared to the degree of stimulation.
- *No central excitatory effect*—there is no central excitatory effect like referred pain or secondary hyperalgesia.
- *Presence of trigger zones*—presence of trigger zones is the most important characteristic feature of true neuralgia.

Painful Neuritis

Neuritic pain occurs as a result of inflammatory influence on pain conduction fibers and is felt in the peripheral distribution of the affected fibers. The inflammatory process alters the relative activity of the fibers that mediate pricking and burning pain elevating the threshold of pricking pain but lowering it for burning pain. This gives the pain of a characteristic burning quality.

Sensory effects such as hyperesthesia, paraesthesia, dysesthesia, and anesthesia may be seen. If motor efferent fibers are affected, then muscular signs such as muscular tic, weakness or paralysis may be seen.

Trigeminal Neuralgia

It is also called as *Tic Douloureux* (painful jerking), *Trifacial neuralgia* or *Fothergill's disease*. The term *Tic douloureux* is only applied when the patient suffers from spasmodic contractions of the facial muscles.

Trigeminal neuralgia is an extremely painful condition as it is unique to humans. It is a syndrome in which symptoms are sufficiently distinctive to permit a reliable diagnosis solely on the basis of history. It is seriousness of these disorders that it has one of highest cause for suicide rates of any disease.

Etiology

- *Dental pathosis*—dental pathosis is believed by some investigators to be involved with the onset of trigeminal neuralgia.
- *Excessive traction*—secondary to excessive traction on the various divisions of the fifth nerve, being influenced by maxillo-mandibular relationship.
- *Allergic*—it can be secondary to an allergic and hypersensitivity reaction causing edema of the trigeminal nerve root.
- *Ischemia*—Wolf thought that ischemia at various portions of the trigeminal pathway might be responsible for the paroxysms of pain.
- *Compression distortion phenomenon*—Jannetta and others have shown subtle changes of a compression-distortion phenomenon which is usually caused by arterial loops of atherosclerotic vessels. Vessels become elongated with advancing age and with atherosclerotic involvement gain abnormal positions by wedging into the space between the pons and trigeminal nerve. It is postulated that with progressive material elongation, fascicles of adjacent nerves later suffer myelin injury and pain results.
- *Mechanical factors*—like pressure due to aneurysms of the intrapetrous portion of the internal carotid artery that may erode through the floor of the intracranial fossa to exert a pulsatile irritation on the ventral side of the trigeminal ganglion.
- *Anomalies of superior cerebellar artery*—it is the most recently blamed cause for trigeminal neuralgia. It lies in contact with the sensory root of the nerve and implicated as a cause of demyelination. Surgical elevation of artery or decompression of the sensory root has high success rate in relieving paroxysmal pain in case of idiopathic trigeminal neuralgia.
- *Secondary lesion*—conditions such as carcinoma of the maxillary antrum, nasopharyngeal carcinoma, tumors of peripheral nerve root, intracranial vascular anomalies, and multiple sclerosis may be presented with trigeminal pain.

Clinical Features

- *Age and sex distribution*—it usually occurs in middle and old age, the disease seldom occurs before 35 years of age. Incidence increases with age due to degenerative changes of the nerve fibers. It most frequently occurs in women.
- *Site*—it is more common on the right side and the lower portion of the face is more frequently affected.
- *Nature of pain*—the pain is paroxysmal, lasting only a few seconds to a few minutes and is usually of extreme

intensity. It may be described by the patient as resembling 'knife like stabs', 'lightening', 'electric shock', 'stabbing' or 'lancinating' type of pain. During the intervals between these violent experiences, there is usually no pain or a mild or dull ache. Attacks do not occur during sleep.

- **Location of pain**—the pain is confined to the trigeminal zone, nearly always unilateral and, if bilateral, is successive rather than concomitant. The mandibular and maxillary divisions are more commonly involved than the ophthalmic. In some instances, these two divisions may be simultaneously affected. The pain never crosses the midline.
- **Aggravating factors**—the pain is provoked by obvious stimuli to the face. A touch, a draft of air, any movement of the face as in talking, chewing, yawning or swallowing may evoke a lancinating attack. Later the pain may be so severe that the patient lives in constant fear of an attack. Often there is a transitory refractory period after the attack. As the attack occurs, the patient may clutch his face as if in terror of the dreaded pain.
- **Triggers zones**—'trigger zones' which precipitate an attack when touched, are common on the vermilion border of the lips, the ala of the nose, the cheeks, and around the eyes (Fig. 18-11). The patient learns to avoid touching the skin over the trigger zones which frequently makes him go unwashed or unshaven for days. Occasionally, only auriculotemporal branch may be involved. The pain and triggering is located in the auricle as well as in the temporal region.

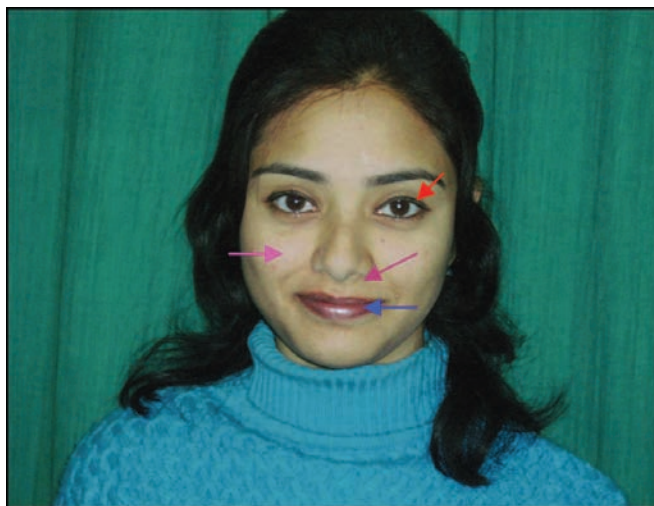


Fig. 18-11: Common trigger zone in patient of trigeminal neuralgia. Lip, ala of nose, cheeks and eyes.

- **Neurological examination**—the neurological examination findings are normal with no objective sensory loss along the trigeminal nerve.

- **Frozen or mask like face appearance**—in extreme cases, the patient will have motionless face—the 'frozen or mask like face'.
- **Associated features**—trigeminal neuralgia may be accompanied by excess lacrimation, conjunctival injection and intense headache.
- **Effect on activities of patient**
 - **Unshaven face**—male patients avoid shaving.
 - **Avoid brushing**—as patient avoids brushing of teeth so oral hygiene is poor.
 - **Poor quality of life**—many patients will lead a poor quality of life, because of the excruciating pain.
 - **Indiscriminate dental extraction**—it is very common for these patients to undergo indiscriminate dental extractions on the effected side without any relief from the pain, because the pain of the trigger zone and pain fiber distributions often mimic pain of odontogenic origin.

Diagnosis

- **Clinical diagnosis**—abrupt onset of pain with trigger point, pain is extreme in nature with less time duration. Pain is localized to known distribution of trigeminal nerve. Spontaneous remission can occur.

Differential Diagnosis

- **Migraine**—a very common condition mistaken for trigeminal neuralgia is migraine or migranous neuralgia ((Horton's syndrome, histamine headache, histamine cephalgia), but this severe type of periodic headache is persistent, at least over a period of hours, and it has no trigger zone.
- **Sinusitis**—pain is not paroxysmal and no trigger zone present.
- **Toothache**—this can be easily identified by examining the oral cavity.
- **Multiple sclerosis**—it occurs because of autoimmune process. Specific clinical features are Charcot's triad, i.e. intention tremor, nystagmus and dysarthria or scanning speech.
- **Tumors of nasopharynx**—tumors of the nasopharynx can also produce similar type of pain, manifested in the lower jaw, tongue and side of the head with associated middle ear deafness. This complex lesion is called as trotter's syndrome. Here the patient exhibits asymmetry and defective mobility of the soft palate and affected side. As the tumor progresses, trismus of internal pterygoid muscle develops, and patient is unable to open the mouth. Here, the actual cause of pain is the involvement of mandibular nerve in the foramen ovale.
- **Post herpetic neuralgia**—neuralgia occurs after attack of herpes zoster virus. Pain is usually involved in the ophthalmic division. The history of skin lesion prior to the onset of the neuralgia usually aids in the diagnosis.

Management

Medical treatment

- *Trichloroethylene inhalation*—it has been proved to be of value.
- *Topical capsaicin cream*—topical capsaicin (nociceptive substance P suppressor) can be applied on affected area.
- *Anti-cholinergic drugs*—it was used for a short period during the late 1960s. Nowadays, this treatment modality is not used.
- *Dilantin*—diphenylhydantoin, an anti-convulsant drug has been recommended, which is effective when given orally, 300 to 400 mg per day. It may be given in single or divided doses. Side effects can include ataxia, mental confusion, nystagmus, gastrointestinal disturbance or skin rashes. Diabetic patients may have hyperglycemia secondary to the therapy and should be closely observed.
- *Carbamazepine* (tegretol) has a special effect on the paroxysmal pain. The use of this drug causes paroxysms to become separated by intervals of freedom for weeks, months or even years. These drugs decrease conductance in Na⁺ channels and inhibit ectopic discharge. This is considered to be the best conservative treatment for trigeminal neuralgia. As an initial dose, 100 mg is given twice daily until relief is established. At no time, the daily dose should exceed 1200 mg. Side effects include dizziness, unsteady gait, gastrointestinal distress, skin rashes and aplastic anemia. Nowadays, another anticonvulsant gabapentin can be given.
- *Baclofen*—recently, baclofen an antispastic drug is also being used.
- *Combination therapy*—a combination of Dilantin and Carbamazepine may also be given.
- *Clorazepam*—an anti-epileptic belonging to benzodiazepine group has been found to be useful.
- *Anti-inflammatory agents*—anti-inflammatory agents like indomethacin and short courses of steroids have been found to be useful.

Surgical treatment

- *Injection of the nerve with anesthetic solution*—local anesthetics injected near the peripheral branches of the trigeminal nerve serves to provide temporary relief from pain and helps in the diagnosis. Some patients get relief after the injection but in some cases, 6 to 7 injections may have to be given to get relief.
- *Injection of the nerve with alcohol*—the most popular material, alcohol, can be placed directly into the area where a nerve exits from the skull or more peripherally. When alcohol contacts the nerve, neurolysis occurs distal to the injection site. Nerve regeneration occurs in 6 to 24 months for most patients. But, the duration of relief from alcohol injection tends to decrease with repeated attempts probably because of the inability of alcohol to

diffuse through the fibrous tissue that resulted from previous injections. Injection of the entire 2nd division at the foramina rotundum and the 3rd division at the foramen ovale may be made in cases where peripheral injection does not give complete relief. If the injection in the ganglion is successful then the anesthesia will last from 6 months to 1 year. Generally 95% alcohol is used or procaine or monocaine 2%, chloroform 5%, absolute alcohol 70%, Ringer's solution 23% can also be used.

- *Nerve sectioning and nerve avulsion (peripheral neurectomy)*—this procedure is more lasting and effective than an injection with alcohol. Nerve sectioning is generally performed on the nerve which cannot be avulsed. The procedure can be performed on lingual, mental or buccal nerve. Peripheral neurectomy results in high degree of success in elimination of pain. But disadvantage of this technique is that result is temporary as nerve may regenerate. Peripheral neurectomies interrupt afferent impulses to the central trigeminal area and also cause damage and degeneration in the gasserian ganglion and thus inhibit nerve impulses from producing pain.
- *Electrocoagulation of gasserian ganglion*—diathermy apparatus is placed in the gasserian ganglion to coagulate and destroy it.
- *Percutaneous radiofrequency trigeminal neurolysis*—it is a technique in which there is use of controlled radio-frequency. It is performed by insertion of temperature monitoring electrode through foramen ovale into trigeminal ganglion. The temperature is raised to 60 to 90 degree Celsius. Advantages include decreased mortality and morbidity and permanent cure. Disadvantages include development of painful anesthesia, keratitis, corneal anesthesia, and sixth cranial nerve palsy.
- *Decompression and compression*—
 - *Percutaneous microcompression*—in this, inflated balloon is used to compress the gasserian ganglion.
 - *Microvascular decompression*—retromastoid craniotomy is carried out and the offending vascular structures are dissected free of the nerves at root entry zone and maintained in that position by insertion of a small piece of gelfoam or Ivalon sponge.
- *Rhizotomy*—actual cutting of trigeminal sensory root results in permanent anesthesia in most patients. The recurrence rate of trigeminal neuralgia after rhizotomy is 20%.
- *Gamma knife radiosurgery*—gamma knife radiosurgery of gasserian ganglion shows success in many patients.
- *Bulbar trigeminal tractotomy*—sensory origin of trigeminal nerve runs from the upper pons through the medulla and in upper cervical cord. This descending tract of the trigeminal nerve may be cut in the area of medulla oblongata to induce loss of pain and temperature sensation with nearly total retention of proprioception.

Glossopharyngeal Neuralgia

It is also called as '*vaguglossopharyngeal neuralgia*'. It is a variant of Tic douloureux that can mimic oral pathologic condition in which pain is confined to the distribution of the ninth cranial nerve.

Clinical Features

- *Age and sex*—this neuralgia occurs without any sex predilection in the middle aged or older persons.
- *Distribution*—the pain is generally unilateral.
- *Nature of pain*—it manifests as sharp excruciating, electric like, lancinating paroxysms of pain in the ear, pharynx, nasopharynx, tonsils or the posterior portion of the tongue. Attacks are of short duration of about 30 to 60 seconds.
- *Trigger zone*—the patient usually has a trigger zone in the posterior oro-pharynx or tonsillar fossa. An important and frequent trigger is the initiation of the act of swallowing.
- *Associated features*—it can be associated with syncope, hypotension, seizures, arrhythmia and excessive salivation can also occur.
- *Progress*—glossopharyngeal neuralgia has a tendency towards remissions and exacerbations. Pain free intervals of seconds, minutes, hours, days, and years are common.

Diagnosis

- *Clinical diagnosis*—pain like trigeminal neuralgia with trigger zone present in tonsil and pharynx area.

Management

- *Topical anesthetics*—patient may get pain relief after applying topical anesthetics to tonsil and side of pharynx on side of pain.
- *Other therapy*—other therapy like carbamazepine, oxcarbazepine, baclofen, phenytoin, lamotrigine and resection of glossopharyngeal nerve can also be initiated.

Geniculate Neuralgia

It involves the intermediate nerve of Wrisberg, an important component of the facial (VII) nerve.

Causes

- *Neuroma and vascular malformation*—pathological involvement of the sensory intermediate nerve root of the VIIth cranial nerve due to neuroma; vascular malformations, etc. are the causes.
- *Zoster infection*—zoster infection of geniculate ganglion canal also cause this type of neuralgia. This is also called as Ramsay Hunt syndrome.

Clinical Features

- *Age and sex*—females are affected more commonly than males. It occurs more commonly in old aged persons.
- *Location of pain*—ear, anterior tongue, soft palate.
- *Nature of pain*—the pain may be felt in the ear with occasional pain in the palate and the tongue.
- *Trigger zone*—triggering is caused by touching the ear.
- *Vesicle*—this can be seen in ear of the patient in case if it is associated with zoster infection.

Diagnosis

- *Clinical diagnosis*—pain in ear, tongue, and soft palate with vesicle formation may give clue to the diagnosis.

Management

- *Topical anesthesia*—topical anesthesia of the external auditory canal may arrest pain in some cases.
- *Steroid*—short course of high dose steroids therapy is useful.
- *Acyclovir*—this is helpful in case of geniculate neuralgia associated with zoster infection.
- *Other drugs*—other drugs like oxcarbazepine, carbamazepine and gabapentin are also useful.
- *Surgery*—if all the above treatment fails then excision of nervus intermedius and geniculate ganglion should be carried out. Microvascular decompression can also be done.

Occipital Neuralgia

Occipital neuralgia occurs due to trauma, neoplasm, and infection of greater or lesser occipital nerve. It is presented as paroxysmal stabbing pain in the occipital region. There is tenderness at superior nuchal line. It is managed by occipital nerve block, corticosteroid and neurolysis.

Post-herpetic Neuralgia

It is discussed in Chapter 31: Viral Infections.

Symptomatic Neuralgia

Neuralgic pain resulting from pathologic lesions is called as symptomatic neuralgia.

Characteristics

- *Atypical manifestations*—such as prolonged or nearly continuous paroxysms of pain.
- *Bridging of paroxysm*—definite bridging of paroxysms with a more continuous aching or burning pain.
- *Bilateral pain*—bilateral neuralgic pain.
- *More nerve involvement*—neuralgic involvement of two or more cranial nerves simultaneously.

- *Sensory manifestation*—neuralgia along with other sensory manifestations such as hypoesthesia, paresthesia, dysesthesia, anesthesia.
- *Motor manifestation*—neuralgia along with muscular weakness or paralysis or other autonomic signs.
- *Failure to respond to therapy*—failure to respond to reasonable therapy.

Eagle's Syndrome (Elongated Styloid Process) or DISH Syndrome

Nowadays, this syndrome is called as *DISH syndrome* (diffuse interosseous skeletal hypertrophy). It is also called as Stylohyoid syndrome, Carotid artery syndrome. Styloid process is originated from temporal bone. It is connected to hyoid bone by stylohyoid ligament. Elongation of the styloid process leads to impingement or compression of adjacent nerves leading to Eagle's syndrome.

Types

- *Classic type*—the classic type occurring after tonsillectomy resulting from surgical exposure of the styloid process. Scar formation will result in pressure on nerve causing the pain while swallowing.
- *Carotid artery syndrome*—the carotid artery syndrome resulting from calcifications of the stylohyoid ligament and elongated styloid process which encroaches the external or internal carotid vessels produce vascular pain.
- *Traumatic Eagle's syndrome*—it develops after fracture of mineralized stylohyoid ligament.

Clinical Features

- *Age*—it is more commonly seen in adults.
- *Sign*—sometime, elongated styloid process can be visualized in pharyngeal area.
- *Symptoms*—pain in the lateral pharyngeal wall and side of the lower face and neck. It may mimic glossopharyngeal neuralgia.
- *Associated symptoms*—associated symptoms include difficulty in swallowing, sore throat, glossodynia, and headache and dull to severe hemifacial pain. Blurred vision and vertigo have also been seen.
- *Characteristic sign*—patients with Eagle's syndrome will characteristically rotate their head slowly to avoid provoking pain. The pain can be demonstrated when pressing the pharyngeal wall against the styloid process and when swallowing or opening the mouth wide.

Radiological Features

- Elongation can be seen radiographically. It is more commonly seen panoramic radiograph. Mineralization is also noticed on the radiograph (Fig. 18-12).



Fig. 18-12: Elongated styloid process seen on radiograph.

Diagnosis

- *Clinical diagnosis*—pain in lateral pharyngeal wall, blurred vision will aid in diagnosis.
- *Radiological diagnosis*—elongated styloid process can be seen radiologically.

Management

- *Topical anesthesia*—application of topical anesthesia will not relieve pain but infiltration of a local anesthetic around the styloid process will provide relief.
- *Surgical*—it is treated by surgical segmentation or resection of the elongated styloid process.
- *Corticosteroid injection*—local injection of corticosteroid sometimes provides relief.

Other Causes of Symptomatic Neuralgia (Fig. 18-13)

- *Trotter's syndrome*—patient complains of neuralgic pain in the mandible, side of head, tongue, and ear. It is associated with middle ear deafness and defective mobility of the soft palate. Trismus of jaw sets in as a result of involvement of internal pterygoid muscle. The cause is tumor of the nasopharynx, starting in the pharyngeal wall involving the eustachian tube.
- *Pterygopalatine fossa syndrome*—this consists of maxillary dental pain, infraorbital and palatal anesthesia, blindness and pterygoid muscle paralysis. The cause is a metastatic tumor to the pterygopalatine fossa.
- *Jacob's syndrome*—this consists of most of the feature of Trotter's syndrome and the Pterygopalatine fossa syndrome plus ophthalmoplegia. The tumor is an intracranial lesion in the middle cranial fossa.

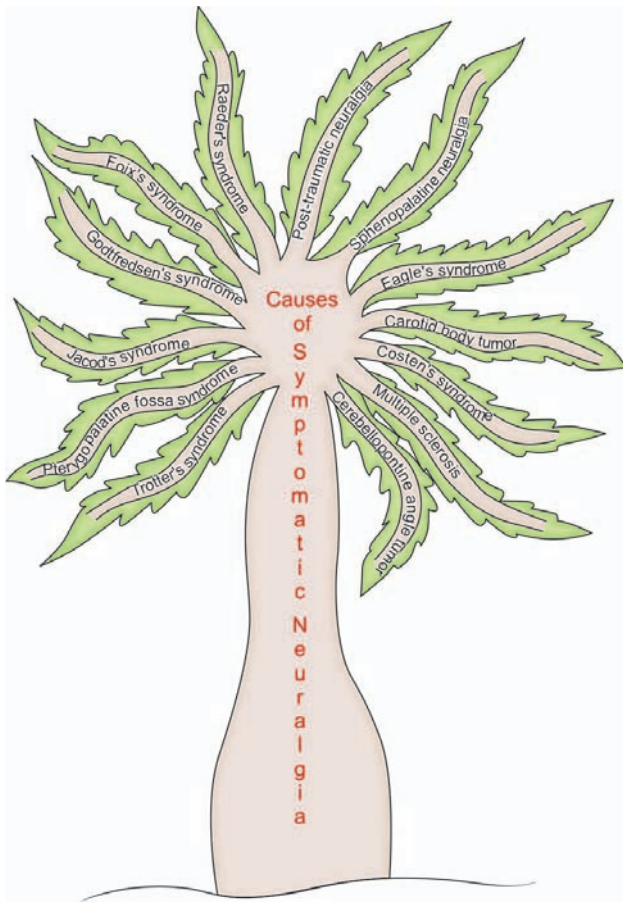


Fig. 18-13: Causes of symptomatic neuralgia.

- *Godtfredsen's syndrome*—it consists of ophthalmoplegia and trigeminal neuralgia usually associated with paralysis of the tongue. The tumor is located in the nasopharynx extending intracranially.
- *Foix's syndrome*—this consists of ophthalmoplegia and maxillary trigeminal neuralgia. This is due to tumor, aneurysm or thrombosis involving the cavernous and/or the lateral sinus.
- *Raeder's syndrome*—it is also called as paratrigeminal syndrome. It is most common in males chiefly those of the middle age and usually appear suddenly. Unilateral severe headache with ocular symptoms like increased lacrimation or greater viscosity of lacrimal fluid, and ipsilateral motor and sensory trigeminal dysfunction. It is due to tumor of middle cranial fossa or of the gasserian ganglion.
- *Post-traumatic neuralgia*—this presents the features which identify it as neurogenous. It combines the characteristics of both painful neuritis and neuralgia. The syndrome has persistent, unremitting though variable, bright, burning pain that suggests painful neuritis and may be accompanied by other sensory, motor and/or autonomic effects that characterize neuritic manifestations.

- *Carotid body tumor*—swallowing dysfunction associated with vocal cord paralysis and patient may complain of glossopharyngeal pain.
- *Costen's syndrome*—it is characterized by impaired hearing, dizziness, and burning sensation in the throat and tongue. Headache about the vertex, occipital region and behind the ears, stuffy sensation in the ears, especially at mealtime, and otalgia.
- *Multiple sclerosis*—it is a disease featuring progressive demyelination of nervous tissue with episodes of relapse and remission. It is slightly more common in females and the usual age of onset is between 20 to 40 years of age. There are a wide range of symptoms reported by the patients with the commonest being : optic neuritis, which is associated with loss of visual acuity and colored vision, and sometimes pain in the eye, weakness of one or both lower limbs, impairment of cutaneous sensations in the lower limbs and trunk, vertigo and ataxia. Bilateral trigeminal neuralgia has been found to occur more in patients with multiple sclerosis than in patients with idiopathic neuralgia.
- *Cerebellopontine angle tumor*—it rarely causes trigeminal neuralgia and pain in the trigeminal area. The majority of these tumors develop within the temporal bone along the course of the vestibular division of the auditory nerve, from where they spread to occupy space between the brain and cranium. The most common symptoms are deafness, headache, tinnitus and loss of balance. Pain, if any, is usually of low intensity, steady, dull ache; hypoesthesia is considerably more common.

Psychogenic Pain

Pain that originates in the mind refers to this pain disorder, for which, there is no apparent physiologic or organic basis and in addition, the patient has a definite history of psychological problems or it implies the presence of emotional or personality disorder.

Psychogenic Intensification Pain

Most psychogenic pains of the mouth and face are of this type. It is usually chronic, lack of adequate peripheral cause, non-anatomical location of the complaint and deviated pattern of behavior raises suspicion about presence of this type of pain syndrome.

Frustration, anxiety, uncertainty, apprehension, fear and tension from the ingredients of pain intensification through the reduced central inhibitory influence on the setting of the synaptic gate in the brainstem.

All conditions, both central and peripheral, that open the gate to the passage of impulses are felt as painful episodes and cause pain that is not due to noxious stimulation; so much is the lack of inhibitory control over the neural impulses that even occurs in the unconscious

state. As a result, teeth may become unduly sensitive or actually painful. They may hurt when touched, moved, pressed, or stressed through occlusal use, or subjected to stimulants such as tooth brushing, flossing, thermal changes or ingestion of sweet. In such situations, many or all of the teeth may become painful. Tension and anxiety may lead to spasm of striated and smooth muscles, inducing local conditions that can become painful.

Conversion Hysteria Pain

Psychic energy from a repressed idea or complex is converted into nervous stimuli giving rise to physical symptoms. Psychological trauma sustained by the patient may become expressed as physical illness. It has been described as 'psychogenic regional pain syndrome'. It is hallucinatory and remotely localized in a way that is psychologically related to the noxious situation.

Delusional Pain

It is created in the subject's mind as a hallucination and is symptomatic of serious psychiatric problems. Delusional pains of the mouth and face is a manifestation of schizophrenic hallucination which is very rare.

Burning Mouth Syndrome

Burning mouth syndrome is common dysesthesia (sense distortion) described by patient as burning sensation of oral mucosa in absence of apparent mucosal alteration.

Etiology

Local factors

- *Contact allergy*—substances in denture material can be allergic like monomer methyl methacrylate, epoxy resin and glycol.
- *Chronic mechanical trauma*—chronic mechanical trauma may occur due to denture, clasp etc.
- *Habits*—oral habits like clenching, grinding, and chronic tongue thrust habit may be causative factors.
- *Infection*—infection with *Candida albicans*, fusospirochetal infection.
- *Xerostomia*—xerostomia due to irradiation, immunological deficiency, systemic causes, medication focal disease.
- *Other*—other factors like TMJ dysfunction, geographic tongue, oral submucous fibrosis, esophageal reflux, angioedema, trauma to lingual nerve, and acoustic nerve neuroma may cause burning mouth syndrome.

Systemic Factors

- *Nutritional deficiency*—vitamin deficiency like pernicious anemia, other B complex vitamins, folic acid, iron

deficiency anemia may be causative factors for burning mouth syndrome.

- *Diabetes mellitus*—this may be one of important causative factors for burning mouth syndrome.
- *Psychological disorders*—depression, anxiety, psychosocial stress.
- *Gastrointestinal problems*—chronic gastritis, chronic gastric hypoacidity may cause burning mouth syndrome.
- *Endocrine disorders*—hypothyroidism and estrogen deficiency.
- *Others*—menopause, diabetes, medication, AIDS, Parkinson's disease, mercurialism can be causative factors.

Clinical Features

- *Age and sex distribution*—it occurs in 5.1% of the general population and patients older than 50 years of age. Females are affected more as compare to males.
- *Site*—tongue is most frequently affected followed by denture bearing areas, buccal mucosa, throat and floor of the mouth.
- *Nature of pain*—pain is present in the morning and it can persist and become aggravated during the day with maximum intensity being in the evening. Half of the patients have continuous pain which may be mild, moderate or severe. Some patients exhibit waxing and waning pattern that occurs several days and week.
- *Associated features*—headache, insomnia, lethargy, decreased libido, mood changes (irritability, depression, and decreased desire to socialize).
- *Aggravative and relieving factors*—aggravative factors include tension, fatigue, and hot foods. Sleeping, eating and distraction reduce intensity.
- *Scaled mouth syndrome*—this is the term used for patient who suffers from burning due to angiotensin converting enzyme (ACE).

Diagnosis

- *Clinical diagnosis*—waxing and waning pattern presented for many days. Pain is mild with increasing intensity throughout the day.

Management

- *Removal of cause*—any cause which is causing the burning mouth syndrome should be removed. For example denture adaptation, allergy control, habit control, consulting.
- *Nutritional supplement*—nutritional supplement like ferrous sulfate 300 mg TDS, iron sulfate 2.5 ml/day IM, cyanocobalamin 250 mg/day, pyridoxine HCl 25 to 100 mg/day, riboflavin 10 mg/day, thiamin HCl 5 to 30 mg/day, folic acid 0.1 to 34 mg/day should be given.

- *Mood altering drugs*—mood altering drug like chlordiazepoxide shows improvement in patients.
- *Other drugs*—other drugs like amitriptyline, clonazepam, alpha lipoic acid, hydrocortisone locally, nystatin oral suspension, ketoconazole, and diphenylhydramine HCl for rinsing.

Therapeutic Modalities of Pain Control

Analgesics

- *Non-narcotic analgesics*—it includes aspirin, acetaminophen and NSAIDs. This drug inhibits generation of pain by blockage of prostaglandin synthesis, especially prostaglandin-E which, via cyclo-oxygenase pathway, forms from arachidonic acid that occurs as part of the inflammatory process by initiating action on the enzyme cytokinase. Prostaglandin is also useful to maintain protective layer of gastric mucosa. That is the reason, after administration of NSAID patient suffers from gastric irritation and gastrointestinal bleeding.
- *Mild pain*—oral administration of 650 mg of aspirin or 200 mg Ibuprofen every four hours is indicated for mild pain. Aspirin has got numerous side effects like ulceration, epigastric distress, nausea, vomiting and increased bleeding time. Aspirin binds irreversibly to platelet cyclo-oxygenase pathway. This prevents platelet production of prostaglandin and thromboxane, substances that are essential for platelet aggregation, thus increasing the bleeding time. For patients, for whom the use of aspirin-like drugs is contraindicated, oral administration of 600 to 1000 mg acetaminophen can be used. Other drugs used are diflunisal, naproxen, and fenoprofen.
- *Moderate pain*—in cases of moderate pain, 400 to 600 mg of ibuprofen can be used. If there is failure to obtain adequate relief, addition of 60 to 90 mg of codeine will increase the opioid effect. In aspirin intolerant patients, 600 to 1000 mg acetaminophen combined with 60 mg codeine or 5 mg oxycodone is generally used.
- *Severe pain*—the onset of severe pain can be suppressed by the use of long acting local anesthetics such as bupivacaine or etidocaine in combination with NSAIDs.
- *Narcotic analgesics*—it contains morphine like agonist, partial agonist and mixed agonist antagonist. These drugs act on CNS receptors to induce peripheral analgesic effect and they depress nociceptive neurons while stimulating non-nociceptive neurons. It is used in managing severe acute pain.
- *Agonist antagonist drugs*—for example: buprenorphine, butorphanol and pentazocine are used to treat moderate to severe pain.

Adjuvant Drugs

Several pharmacological agents are useful for palliative and cause-related therapy.

- *Analgesic balms*—it gives soothing palliative relief from inflammatory pain of both superficial and deep somatic category when applied topically on exposed or ulcerated area. Aloe Vera juice is an ancient variety used for superficially generated pain. Balsam of Peru and eugenol are well known balms. They are applied in various forms like ointment, liquid, and adhesive dressings.
- *Anticonvulsants*—phenytoin has capability of suppressing pain of paroxysmal neuralgia in about 20% of cases. Carbamazepine which can be used in trigeminal neuralgia is effective in 70% of cases.
- *Antidepressants*—some patients with vascular pain and chronic pain may respond to antidepressant medication. Some antidepressants like tricyclic and MAO (monoamine oxidase) inhibitors are helpful in managing chronic pain syndromes.
- *Antihistaminic*—antihistaminic counteract the vasodilator effect of histamine by blocking certain histamine receptors. They may be useful in allergic conditions and some vascular pain syndromes. A direct analgesic effect is reported by use of several antihistaminics like diphenylhydramine.
- *Antimicrobials and antiviral agents*—antinociceptive benefits of such agents related to resolution of infection that cause pain.
- *Dietary supplements*—the chief relationship between diet and pain appear to be with L-tryptophan which converts into serotonin. Brain and spinal cord serotonin neuron are actively involved in nociceptive response. The synthesis of brain serotonin is dependent on deposition of plasma tryptophan, 90% of which is bound to albumin and must compete with other amino acids at blood-brain barrier. The use of tryptophan and other amino acids as dietary supplement appear to be a valid approach to otherwise refractory chronic pain problems. Adequate dose of 4 mg / day with low protein, low fat, high carbohydrate diet should be given.
- *Neurolytic agents*—the most commonly used is 95% ethyl alcohol, although effective, neurolysis does not produce regeneration of peripheral axons and gives only temporary relief. The hazards include extensive local fibrosis and deafferentation effect restrains its use.
- *Nor-epinephrine blockers*—in analgesic, blocking of the stellate ganglion for the control of causative pain about orofacial region norepinephrine blockers are used.

- *Tranquilizers*—the major tranquilizers like phenothiazine are very useful in pain control by reducing modulatory effect, anxiety and apprehension.
- *Muscle relaxant*—medication that tends to relax skeletal muscle has some value in the treatment of myogenous pain especially. They are helpful in obtaining occlusal disengagement.

Cause Related Therapy

Many who experience orofacial pain have organic structural cause for it. You have to identify the cause and eliminate it. Therapy is given accordingly.

Relaxation

To an extent, tension and stresses are etiological factors in pain. Following techniques are used to provide relaxations.

- *Autosedation*—it has been demonstrated that some functions are automatic and beyond the voluntary influence. However, it can be manipulated by conscious mental procedure. There are a number of relaxation techniques that involve mental discipline and self-training like yoga. Modulating effects of conscious mental autosuggestion is very reactive and should be actively utilized in pain management. Sometimes music may be helpful in relaxation techniques.
- *Biofeedback*—EMG feedback has been found to be useful in reducing hyperactivity of masticatory muscles in individuals who are prone to clench their teeth during day time. EMG biofeedback apertures include pair of surface electrodes made up of silicon rubber, voltmeter with amplifier and visual and auditory display. In training muscle relaxation, the electric potential of masticatory muscles can be recorded and displayed on digital voltmeter so that patients can observe the amount of tension and try to relax. EMG feedback training aims at teaching the patient to become aware of the tension in the masticatory muscle and to learn to relax it.
- *Occlusal disengagement*—occlusal disengagement induces substantial beneficial effects on discomfort of TMJ disorders. Disengagement sets occlusion at rest and causes the masticatory system to relax. Occlusal disengagement can be accomplished by
 - *Voluntarily disengagement*—patient is asked to voluntarily leave its teeth apart.
 - *Chewing gum*—variety of simple habit training as by placing thin parts of chewing gum between molar teeth.
 - *Muscle relaxant*—we can also use muscle relaxant for the occlusal disengagement.
- *Interocclusal device*—interocclusal device significantly decreases nocturnal EMG activity or bruxism. They tend to normalize the sensory and proprioceptive impulses generated by occlusal dysfunction and thus shut off afferent input that initiate muscle splinting and aggravate myoplastic activity. Device uses are disengaging splint, occlusal correcting splint and mandibular re-positioning splint.

Counseling

Armed with good understanding of nature of pain and how it is modulated, the doctor can use counseling as a powerful tool in the management of pain.

- *Emotional stress*—due to anxiety and fear, a person in pain may be emotionally upset which accentuates his suffering. An accurate explanation of pain should be given to the patient in a language that can be understood by him. It will boost the patient's confidence and hope. Honest words of assurance can allay fear and diminish anxiety thus reducing the level of suffering.
- *Evaluation of consequence*—apprehension concerning the consequent damage and expecting future threat to one's well-being cause psychological intensification of painful experience. This can be reversed by open discussion on realistic consequences that exist.
- *Inhibitory influence*—the excitatory influence of attention directed towards the complaint can be countered with distraction by directing the mind towards thinking that is more positive.
- *Coping strategies*—attention and tender loving care should reward illness behavior. Severe pain may be made tolerable by replacing the suffering with something better to do like doing something that benefits others, activities that direct attention away from self or constitute occupational therapy.
- *Hypnotherapy*—anti-nociceptive methods can be used, provided that the pain is not due to psychoneurosis. Some suffering is too valuable to the patient to be given up. Personality, disintegration may follow withdrawal of pain. Effects depend on susceptibility of the patient to suggestion. Hypnosis is effective in large number of patient's presenting symptoms of severe pain of organic origin.
- *Psychotherapy*—it is needful in mainly chronic pain syndromes like conversion hysteria and delusional pain. Pain behavior may be unlearned by manipulation of consequences such as withdrawing positive reinforcement and rewarding better behavior. It requires full knowledge and cooperation not only of the patient but also of those in close personal contact which includes family members, doctors, nurses and the therapist.

Physiotherapy

- **Massage**—it is primarily used to improve biomechanical functioning. It is indicated for use in pain management. Deep massage especially of muscles is an important therapeutic measure in pain management.
- **Exercise**—an immobilized muscle not only loses its strength by disuse atrophy but also shortens due to myofascial contraction. Exercise increases the pain threshold. Forceful contraction of antagonist muscles causes reflex relaxation of agonist muscles. This principle is used in masticatory myospasm. For example, spastic elevator muscle is relaxed by opening mouth against resistance; retrusion of protruded mandible against resistance leads to relaxation of spastic lateral pterygoid muscles. Stimulation of muscle proprioceptors to normalize the excessive EMG activity in painful spastic skeletal muscles can be helpful.
- **Deep heat therapy**—physiotherapy in the form of penetrating heat has its value in treating the patient.

Diathermy is used for treating inflammatory type of pain.

- **Trigger point therapy**—following method is carried out in trigger point therapy.
 - **Spray and stretch method**—in this, technique vapocoolant (fluorocarbon), spring capped bottle with calibrated nozzle that delivers fine stream is needed. The muscle should be moderately stretched just short of pain. The vapocoolant is applied by using parallel sweeps in one direction traveling towards the reference area. The nozzle is held 15 to 18 inches away from the skin and stream is directed at an acute angle of 30 degrees. After 2 to 3 sweeps muscle should be rewarmed. At the end, moist heat is applied and range of motion exercise instituted.
 - **Ischemic compression**—in this, 20 to 30 lbs of pressure at trigger site for 1 minute is applied while the muscle is stretched just short of pain.
 - **Ultrasound therapy**—frequency used is about 27 MH lasting for 10 to 15 minutes. Analgesic effects or short wave diathermy is supposed to be caused partly by mechanical vibration and partly by deeply penetrating thermal energy. The increase in temperature of deep tissues may become considerable (41 to 45 degree) and result in increased elasticity and plasticity of tissue collagen.
- **Physical activity**—in patients with chronic pain, maintenance of the physical activity is important. Extending the exercise to the limit of pain tolerance is significantly beneficial. The more the exercise is performed the fewer is the pain behavior display.

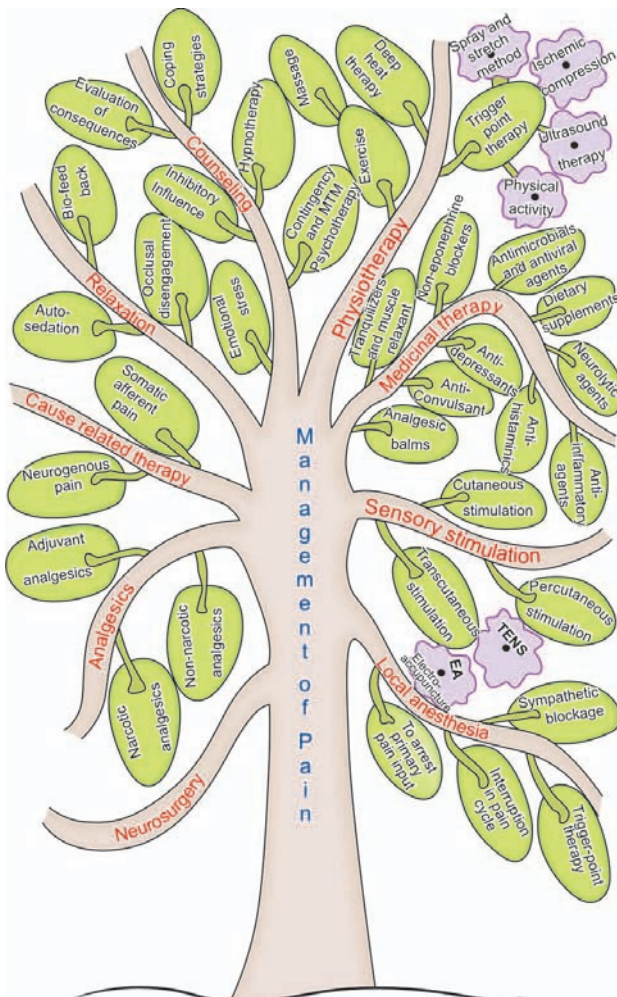


Fig. 18-14: Management of orofacial pain diagrammatic representation.

Sensory Stimulation

- **Cutaneous stimulation**—in this, effects occur through stimulation of thick myelinated cutaneous afferents like beta neuron chiefly. It is done by pressing and rubbing the skin over the site of injury. Superficial massage is also important in reducing pain. It is enhanced by adding stimulating substances like alcohol or menthol ointment. Other methods which used for cutaneous stimulation are use of mustard plaster and circulatory bath water. Mustard plaster is an age-old pain remedy. It is well known that mild stimulation of nociceptor also increases the pain inhibitory mechanism. Circulatory bath water has a therapeutic effect and a brisk stream of shower water directed against neck and back gives relief.
- **Transcutaneous electrical nerve stimulation**—Nowadays, it has become a popular form of pain control. The unit employs low intensity tardic current at a high frequency of 50 to 100 Hz, applied to skin through electrodes attached by a conduction paste. It is used to stimulate nonnociceptive cutaneous afferent neurons that activate descending pain inhibition mechanism without

involving opioid peptides. Analgesic effect ranges from 50 to 70%.

- *Electro-acupuncture*—it utilizes low frequency 2 Hz but high intensity current. It is applied at a specific point called as acupoint. It is used to stimulate muscle nociceptors which in turn activates the endogenous anti-nociceptive effect. The anti-nociceptive effect is not immediate but requires induction period of 15 to 20 minutes. Analgesia may be segmental or general. The segmental relief of pain is thought to involve cerebrospinal fluid enkephalin level while general effects appear to involve the action of beta endomorphin secreted by pituitary gland in the bloodstream.
- *Percutaneous stimulation*—it is done by electrode that penetrates the skin. Subcutaneous nerve stimulation by an electric current produces prolonged anesthesia that is not reversed by naloxone, which indicates that it does not recruit the opioid peptide. New term is percutaneous stimulation of the periosteum by insulated needle. It uses 9 to 12 volts at 100 to 300 Hz for 45 minutes.

Local Anesthesia

- *Analgesic blocking*—to arrest the pain of myospasm by analgesic blocking has marked therapeutic effect. It is preferable to block nociceptive pathway than muscle proper.
- *Interruption of pain cycle*—on the basis of clinical evidence, a cycling mechanism appears to be implicated in several pain syndromes that occur in the orofacial region. Such cycling is effectively interrupted by local anesthetic blockage of nociceptive impulses at the primary source or somewhat along its mediating pain pathway. Remission of pain outlasts the period of anesthesia.
- *Trigger point therapy*—injection of short acting local anesthetics of low myotoxicity at myofascial trigger point located in muscle tissue effectively resolves the referred pain syndrome. This therapy is required when triggered point are located in the muscle tendon.
- *Sympathetic blockage*—the diagnostic analgesic blocking of afferent sympathetic pathway to identify mediation by such neuron. Stellate ganglion analgesic block is as effective as amitidine for treatment of reflex sympathetic dystrophy. Other syndromes in which it is effective are herpes zoster and post-herpetic neuralgia.

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19

Infections of Oral Cavity

Introduction

Facial infections are relatively a common presentation to both, general medical and dental practice. Most originate in superficial structures (skin, subcutaneous tissue, etc.) and are often easily diagnosed and treated.

Infections originating in deeper structures can be severe, rapidly progressive and may cause prolonged morbidity, long-term complications as well as potentially endanger life. Efficient treatment requires accurate diagnosis, early aggressive medical treatment and in most cases, urgent decisive surgical management.

Infection is a clinicopathological entity involving invasion of the body by pathogenic microorganisms and reaction of the tissues to microorganisms and their toxins. Soft tissue infections of head and neck are commonly encountered in routine practice in dentistry. These infections may be odontogenic or non-odontogenic in origin. Once the infection extends past the apex of the tooth the pathophysiology of the infectious process can vary, depending upon the number and virulence of the microorganisms, the host resistance and anatomic geography.

These odontogenic infections can become severe, life threatening facial space infections. These infections have the potential to spread through facial planes of head and neck thereby compromise the vital structures in these regions, e.g. intracranial odontogenic infection leading to necrotizing fascitis of head and neck. Most odontogenic infections can be managed successfully with minimal complications. The key to successful management is sound surgical principles.

Host Defense Mechanism

The relationship between the host and microbes is a dynamic one. Usually, host resistance is the dominant factor. On the other hand, when the host resistance is

lowered, microbes predominate and clinical infection occurs. In establishing the presence of infection there is interaction between three viz. factors host, environment and microbes.

A compromised patient is more likely to have infection and this infection can rapidly acquire a serious form. Hence the patient's history is one of the important criteria to recognize the patient ability to defend himself against the infection. The adverse relationship between the host and the infectious microorganism can be best understood by imagining a balance on which the pathologic attribute of microbes are weighed against the protective mechanisms of the host.

Body defends against the microbial invasion by three major defenses—local defense, cellular defense and humoral defense. The microorganisms on the other hand use two weapons in this battle, i.e. virulence and number of microbes.

Local Defense

- *Epithelial lining*—it is the first line of defense. It physically hinders the penetration of surface bacteria into deeper tissues. When there is a break in the continuity of the anatomic barriers, the microbes find an easy way into deeper tissues. In the oral cavity, the bacterias find way through deep periodontal pockets and necrotic dental pulp.
- *Secretory and drainage system*—this system assists host defense by physical and chemical action. The mucociliary activity, peristaltic motion and flushing action, that all result in drainage and mechanical removal of bacteria. Obstruction and impairment of drainage almost always result in infection. Secretion of the oral cavity, i.e. saliva and swallowing mechanism helps to remove the food particles and also prevents stagnation of microbial flora at one site. Chemical constitution of saliva changes as the pH changes. This favors the growth

of bacteria. Hence, the pH of saliva should be maintained to control the bacterial growth.

- *Microbial floral interference*—this refers to inhibitory effect exerted by one microorganism on the growth and population of other. The normal flora of oral cavity is able to discourage the growth of new species of bacteria.
- *Mucosal immune system*—beneath the mucosal epithelium, lamina propria and connective tissue contain a large number of immunocompetent cells. The B lymphocytes and plasma cells with the help of certain T lymphocytes locally synthesize IgA, some IgE and small amounts of IgG and IgM. The immunoglobulin in secretion together with the local protective factor constitutes mucosal immune system. IgA inhibit the colonization of mucosal epithelial cells by occupying potential sites of attachment. They also neutralize the toxins and viruses.

Humoral Factor

- *Immunoglobulin*—the host synthesizes specific protein molecules, with antibody activity, in response to antigenic stimulation. These proteins are derived from B lymphocytes and plasma cells and are called as IgA, IgM, IgD, IgG and IgE. Secretory IgA and IgE antibodies directly prevent the attachment of pathogenic bacteria or parasites to epithelial receptor cells, which is very crucial in preventing colonization and subsequent infection. By direct action, antibodies can neutralize viruses and microbial toxins.
- *Complement system*—this consists of group of serum proteins, which by a series of reactions produce and release byproducts whose functions are to initiate inflammatory reaction, regulate and enhance phagocyte function and attack the bacterial cell membrane. The activation of complement system by either pathway results in generation of cytolytic activity, chemotactic factor and mediator, anaphylactic toxin, opsonising and phagocyte enhancing activity, and immune adhered activity.

Cellular Component

- *Polymorphonuclear leukocytes*—PMNs are the first phagocytes to respond to the chemotactic factor elaborated by complement system. Thus, they predominate in acute infection, which persists for about one week. Their function is to clear up the battle field.
- *Lymphocyte*—it consists of two types of cells—B lymphocyte and T lymphocyte. Both T cells and B cells produce lymphokinin hormone that plays several vital roles in resistance to infection. Primarily they regulate the action of phagocytic cells. Thus lymphocytes are responsible for modulation and control of phagocytic cells.

- *B lymphocytes*—they are responsible primarily for combating extracellular pathogens, particularly those bacteria that are not able to survive inside phagocyte or other cells which secrete specific antibodies against which, the host has previously been exposed.
- *T lymphocyte*—T cells, on the other hand, are responsible for combating intracellular pathogens which are primarily viruses and those bacteria that can survive within phagocyte cell. In addition, T cells are responsible for surveillance against the proliferation of tumor cells. These complex functions of T cells are described in general as cell mediated immunity.
- *Macrophage inhibiting factor*—it inhibits the random movement of macrophages away from the site of infection, keeping them where they are needed.
- *Interferon*—it enhances lysis of bacteria by phagocytes.
- *Mitogenic factor*—secreted by T cells; stimulates B cell proliferation in the body. This will result in antibody response to antigenic stimulation.

Microbial Factors

Pathogenicity

- *Definition*—pathogenicity is the ability of a microbial species to produce disease.
- *Mechanism*—it involves subversion of host cell metabolism, and production of host cell lytic factor.
- *Toxin*—production of potent toxins which systemically act on the host. To set up an infectious process, a microorganism must be able to do the following:
 - To enter the host
 - To multiply in host tissues
 - To resist host defense
 - To damage the host.

Virulence

- *Definition*—it is the term applied to the properties in particular strain microorganisms. Virulence is the sum total of several determinants such as adhesion, toxigenicity and communicability.
- *Exaltation and attenuation*—enhancement of virulence is known as *exaltation* and reduction of virulence is known as *attenuation*.
- *Adhesion*—the initial event is the attachment of bacteria to the body surface. This attachment is not a chance event, but it is a specific reaction between the surface receptors on the epithelial cells and the adhesive structures on the surface of the bacteria.
- *Invasiveness*—this refers to the ability of a pathogen to be spread in the host tissue after establishing infection. Highly invasive pathogens cause generalized infections and less invasive one causes localized infections.

- **Toxigenicity**—bacteria produce two types of toxins, i.e. exotoxin and endotoxins. Exotoxins are heat labile proteins excreted by certain bacteria and they diffuse readily into the surrounding tissues. Endotoxins are heat stable polysaccharide protein lipid complexes, which form an integral part of the cell wall of gram negative bacteria.

Effect of Infection on Host

- **Inflammatory reaction**—infectious agents initiate, in the host, a series of reactions that are collectively called as 'inflammatory reaction'.
- **Protective reaction**—this response results in generation and release of mediators, microvascular changes and mobilization and activation of leukocytes, all designed to eliminate the infectious pathogens and repair tissue injury. Therefore, these reactions are protective in nature.
- **Direct injury to host cells**—direct injury to the host cells, enhancement of the parasite's invasiveness and amplification of these effects occur due to neutralization of host defence.
- **Symptoms**—there is fever, endotoxic shock and intra-vascular coagulation.
- **Hypersensitivity reaction**—the pathogen or its products, in certain situations, may combine with antibodies or sensitized mononuclear leukocytes to produce harmful immunologic effects called as 'hypersensitivity reaction'.
- **Compromised host**—person whose defense mechanisms have been lowered as a result of diabetes, tuberculosis, rheumatic fever, malignancy, radiation therapy, use of therapeutic immunosuppressive drug or antibiotics, extensive skin burns, genetic deficiency of immune system and malnutrition.

Oral Microflora

The microbial flora that is characteristic of a particular site in majority of the population is referred as the indigenous flora or normal flora. The oral microflora at birth is aerobic and after eruption of teeth, anaerobes are also established. The intrinsic source of microorganisms in the oral cavity is gingival crevice material, pus cells and epithelial cells undergoing degradation and salivary component.

Anatomic Considerations in Dentoalveolar Infection

- **Maxillary central incisor and lateral incisor**—the apices of the roots of central and lateral incisors lie closer to the labial alveolar process. The infection from these therefore exits through the bone in this region. The further spread is influenced by orbicularis oris and the subcutaneous tissues at the base of nose, which tend to limit the infection at the base of the nose.
- **Maxillary canine**—the position of the roots of the canine is in the labial aspect. So, the infection will exit from the bone on the labial aspect. The relation of the levator anguli oris muscle then determines whether the infection will be localized in the oral vestibule or will progress extra-orally. If the site of perforation is below the attachment of the levator anguli oris, then vestibular swelling will occur. If the site of perforation is above the muscle attachment, infection enters the canine space.
- **Maxillary premolar**—infection from the roots of premolars generally exits from the bone on buccal aspect and occasionally, on the palatal aspect. Because of the muscle attachments in the buccal region (zygomaticus major and minor and levator labii superioris), which are situated above the root apices of upper premolars, infection from these teeth will tend to localize within the oral vestibule. On the other hand, sometimes due to long roots of premolar, a canine space infection may develop.
- **Maxillary molars**—periapical infections from upper molars usually perforate the buccal aspect of the alveolar process, but on occasion, a palatal abscess may be encountered. The relationship of the attachment of buccinator muscle is the factor that determines whether localization is intraorally or extraorally. If the perforation occurs below, muscle swelling will appear intraorally and in reverse situation, infection extends laterally to buccinator muscle and form buccal space abscess.
- **Mandibular incisor**—it will reach labial aspect of the alveolar process. In this region, relationship of mentalis muscle to root apices of these teeth determines the further course of infection. If the infection breaks through the bone above mentalis muscle, swelling will appear in the vestibule. If the infection breaks through the bone below the attachment of the mentalis muscle, swelling will appear extraorally. It may remain localized in the subcutaneous tissues of chin or it may spread into the submental region.
- **Mandibular canine**—as the muscle attachments in the region of mandibular canine are located well below the apex of the tooth, periapical infection from this tooth will localize in the oral vestibule, after extending through the labial cortical plate.
- **Mandibular premolar**—infections from lower premolars penetrate the buccal cortex. The further extension is governed by depressor labii inferioris, depressor anguli oris and platysma. This results in vestibular abscess. Rarely, lingual cortical perforation may occur and result in sublingual abscess.

- *Mandibular first molar*—infection from lower first molar can also drain to buccal aspect of bone, below the attachment of the buccinator. The oblique line of buccinator muscle attachment on mandible, however, generally results in root apices being above the origin of this muscle thereby causing localization of infection within the oral vestibule.
- *Mandibular second molar*—due to the position of second molar in the alveolar process, there are 50% chances of perforation of the infection either buccally or lingually. There are equal chances of the root apices to be either above or below the attachments of the mylohyoid or buccinator. Hence there are 4 possible sites for localization of infection arising from these teeth:
 - On the buccal aspect abscess may either be form in the buccal vestibule.
 - It may appear in the buccal space.
 - On the lingual surface, exit of infection above mylohyoid will result in sublingual abscess.
 - Perforation below the mylohyoid muscle results in submandibular space involvement.
- *Mandibular third molars*—it is generally positioned medial to the vertical plane of ramus. Therefore, its apex is much closer to the buccal cortical plate. In this region, attachment of mylohyoid muscle is near the alveolar margin and its posterior border is just behind the tooth. Due to this relationship, infection from vertically positioned third molar will tend to involve the submandibular space. With mesioangularly and horizontally placed teeth the infection will tend to spread beyond the posterior extent of mylohyoid muscle and localize in pterygomandibular space. Occasionally, infection from third molars can involve the sub-masseteric space.

Spread of Infection

Direct Invasion or Extension (Table 19-1)

It is the propagation of an infection between the layers of fascia; from one area to fascia of another area. It can occur secondary to distension and pressure on the body. This will force contents of infection through facial layers by hydrostatic pressure.

- *Spread of infection within the bone*
 - *Alveolar bone*—the bone of the alveolar process is quite similar to the dental structure, in terms of response to infection. The alveolar bone consists of interconnecting marrow spaces delimited by unyielding calcified tissue, all of which are enclosed circumferentially by layers of cortical bone of varying thickness.
 - *Invasion of bacteria in marrow*—the invasion of bacteria from marrow space triggers the process of inflammation and cause some consequences of edema,

Table 19-1: Sites of localization of acute dental infections

Involved teeth	Usual exit from bone	Relation of muscle attachment to root apices	Site of localization
Upper central incisor	Labial	Above	Oral vestibule
Upper lateral incisor	Labial palatal	Above	Oral vestibule Palate
Upper canine	Labial	Above Below	Oral vestibule Canine space
Upper premolars	Buccal palate	Above	Oral vestibule Palate
Upper molars	Buccal Palatal	Above Below	Oral vestibule Buccal space Palatal
Lower incisors	Labial	Above Below	Submental space Oral vestibule
Lower canine	Labial	Below	Oral vestibule
Lower premolars	Buccal	Below	Oral vestibule
Lower first molar	Buccal Lingual	Below Above Below	Oral vestibule Buccal space Sublingual space
Lower second molar	Buccal Lingual	Below Above Below Above	Oral vestibule Buccal space Sublingual space Submandibular space
Lower third molar	Lingual	Above	Submandibular or pterygomandibular space
Involve teeth	Usual exist		

ischemia, necrosis and isolation from systemic circulation and immune system. This is the process by which bacteria survive within the bone of jaw.

- *Spread of infection*—the path of least resistance is along the medullary spaces. This explains the ability of odontogenic infection such as osteomyelitis, to spread for great distances along the jaw before they erode through the cortical plate. Another factor in the spread of infection within the bone is the thickness of the cortical plate on either side.
- *Spread in maxilla*—in maxilla, the buccal cortical plate is thinner as compared to the palatal; hence the abscesses erode through the buccal cortical plate.
- *Spread in mandible*—in mandible the buccal plate of bone over the bicuspid and molar is thick, while the lingual plate is thinner; hence the infection from lower

molar teeth spread lingually, eroding through the lingual cortical plate and then spreads into the soft tissues of floor of mouth.

- *Spread of infection within the soft tissues*
 - *Muscle and fascia*—muscle and fascia function as anatomic barriers to spread of infection within the soft tissues because muscles have tight dense capsule surrounding them and are well vascularized. They are more susceptible to bacterial invasion than loose connective tissue surrounding them. Muscles and fascia of head and neck are having potential space within which the invading bacteria can propagate for a while. This contain various organs including nerves and blood vessels, salivary glands, lymph nodes and fat with surrounding loose fibrous connective tissue. These are the pathways around which infections can spread.
 - *Stage of inoculation*—if the infection does not perforate the oral mucosa, it will enter the fascial spaces of head and neck region. For several days after their initial entry into the soft tissues the bacteria will grow without triggering an intense inflammatory response. A mild edema may be present in the affected area and the patient may experience soreness. This minor swelling can later become soft and doughy on palpation. This stage of infection responds quite well to either extraction or endodontic treatment of the offending tooth.
 - *Stage of cellulitis*—as the bacteria that have inoculated the soft tissue space multiply, they elaborate toxins and metabolic byproducts due to which, an intense inflammatory response is triggered.
 - *Stage of abscess*—as the cellular phase of inflammation progresses, inflammatory cells consisting of many polymorphonuclear leukocytes are drawn to the area of infection by various lymphokines, including leukotoxin. These phagocytes engulf the bacteria and digest them. Other lymphokines cause necrosis of the surrounding soft tissues.
 - *Resolution*—one or two days after the beginning of infection, specific immune system consisting of antibiotics and activated T lymphocytes come into play. This is called as ‘round cell infiltration’, consisting of lymphocyte macrophages and plasma cells. Macrophages are efficient in phagocytosis of bacteria and necrotic debris and are able to dispose off this material.

Spread by Lymphatic System

The face and neck infections result in regional lymph node enlargement, which results from proliferation of lymphocytes and histiocytes in response to antigenic stimulation.

If the antigen is catabolized and becomes non antigenic, the process of lymphocyte activation ceases and the nodes will return to their previous size.

Spread by Blood Vessels

The hematogenous spread of head and neck soft tissue infection may result in very serious complications such as bacteremia, resulting in distant infections involving the heart valve, and cardiac, vascular and orthopedic prosthesis. The microorganisms entering the vascular system may also result in thrombophlebitis, problems particularly involving the intracranial sinus, e.g. cavernous sinus thrombosis. Another serious consequence is generalized septicemia, which can result in shock and death.

Diagnosis of Odontogenic Infection

To arrive to an accurate diagnosis and for appropriate treatment, the basic principles of patient evaluation must be firmly observed. Proper patient evaluation is based upon:

- *Careful history including review of all systems*—the clinician should obtain history of the present illness as it relates to the onset, history of toothache and headache, nature, location and duration of pain, chills, etc.
- *Physical examination*—a comprehensive regional examination of the patient must include inspection, palpation and percussion. Size of the swelling and its relationship to the anatomic structures should be noted on inspection. Palpation is used to confirm the size, note tenderness, temperature and to assess regional lymph nodes and fracture. Percussion with metallic instrument is useful in determining periapical infections.
- *Appropriate laboratory studies*—sometimes a clinician may need to establish a diagnosis by laboratory confirmation, prior to any treatment. The investigations required while dealing with odontogenic infections may be complete blood count, blood sugar level, ESR, gram staining, culture and sensitivity, Montoux test, blood culture, biopsy, radiograph and computed tomography.
- *Proper interpretation of findings*—all these findings, complete history and examination should be properly interpreted then and diagnosis should be made.

Pathophysiology of Infection

- *Cardinal sign*—the body’s response to infectious agent is inflammatory, which is essentially a protective phenomenon. Hence, the cardinal signs of inflammation are present, to some degree, in nearly all patients with infection.

- *Redness (rubor)* is seen when the infection is close to the tissue surface, which is secondary to the intense hyperemia caused by increased vasodilation of arterioles.
- *Calor or heat* is due to inflow of relatively warm blood from deeper tissues, increased velocity of blood flow and increased rate of metabolism.
- *Dolor or pain* results from pressure on sensory nerve endings, from distension of tissues caused by edema or spread of infection. Release of substance like kinins, histamines or bradykinin is also responsible for pain. It is the most universal sign of infection.
- *Swelling* accompanies infection, unless the infection is confined to bone which cannot swell. It is due to the accumulation of fluid, exudate or pus.
- *Loss of function* is another sign of infection. A patient immobilizes the painful part in the most comfortable position he can find. Hence, when the masticatory muscles are involved, there is limitation of jaw movement.
- *Fever* occurs in some cases, which reflect a non-specific physiologic response of host to tissue injury. This injury results in increase of substance called pyrogen from endogenous (injured tissue) and exogenous source (infecting agent). In clinical fever, it appears that the hypothalamic regulating center is stimulated by endogenous pyrogen, which is activated by bacterial endotoxin release from granulocytes, monocytes and macrophages.

Principles of Management of Odontogenic Infection

The primary treatment of odontogenic infection does not end with extraction of offending tooth. It also includes establishment of adequate drainage and administration of appropriate antibiotics along with maintenance of airway, when indicated.

- *Proper assessment of airway*—often this becomes difficult due to limited mouth opening, which frequently accompanies these facial space infections. To assess it properly, mouth opening should be measured; tongue mobility and elevation of floor of mouth should be checked lateral pharyngeal edema and deviation of uvula should also be checked. If the airway compromise is recognized early, then an awake fibro-optic laryngoscopy or blind nasal intubation should be used. If intubation can not be performed while the patient is awake, tracheostomy should be considered.
- *Evaluation of vital parameters*—pulse, temperature and respiration should be recorded. If temperature is more than 103 degree Fahrenheit, then it is a cause of concern. In such cases, besides employing local and systemic

measures of lowering the temperature, culture of blood and urine should be done to check for septicemia and a radiograph must be taken for checking aspiration pneumonia.

- *Hydrate the patient*—most of the patients have poor oral intake for several days, secondary to dysphagia and pain. Fever also increases the metabolic requirements of a body. Intravenous hydration should be initiated and monitored by normalization of vital signs and urine output. Along with IV fluids, rich nutritional supplements can be given to improve the nutritional status.
- *Good clinical examination*—good clinical examination with proper history is very important. While taking history, assessment of pre-existing systemic conditions such as diabetes mellitus, tuberculosis should be done.
- *Empiric antibiotics therapy*—with serious infection it is often necessary to begin antibiotic therapy before the culture result is available. This is called as empirical therapy which is directed towards organisms most likely to cause that infection. Most often the empirical antibiotics of choice for oral infection are penicillin and for penicillin allergic patients, erythromycin should be given.
- *Incision and drainage*—it relieves pain and hastens resolution and healing. Ideally an abscess should be incised at its earliest sign of ripening, i.e. when only thin budging mucosa separates the scalpel from the infection. The incision should be located at a cosmetically and functionally acceptable place, such as under the shadow of jaw line or in a hair bearing area. Intraorally, the incision should be avoided at sites of oral function, such as labial frenum or depth of vestibule where the denture border may rest.

Sequelae of Pulpitis (Pulpopariapical Abscess)

- *Causes of pulpitis*—causes for pulpitis is due to irritating inflammatory products. Various factor like host resistance, number and virulence of bacteria, amount of antigenic material, and degree of tooth function and extent of other trauma also play a vital role.
- *Local inflammatory response*—these products escape from pulp canal and initiate local inflammatory response. The pulp is non-vital in such cases.
- *Periapical lesion*—if none of the irritating products of pulp necrosis reach the periapical tissue, then no periapical pathosis is induced. If infection reaches the periapical tissue then in some cases host defense effectively combats and localizes the resultant inflammation in circumscribe area.
- *Acute periapical abscess*—teeth with contaminated gangrenous pulp have large number of bacteria passing

in the root canal which may be sufficient to overwhelm the defence of periapical tissues resulting in an acute periapical abscess.

- *Chronic alveolar abscess*—resultant infection may be partially controlled by body defence, by surgically induced draining or by antibiotic therapy, thus results in the development of chronic periapical periodontitis or chronic alveolar abscess.
- *Radicular cyst*—odontogenic epithelial rests of Malassez (present in PDL and periapical granuloma) proliferate a radicular cyst may develop in response. If adequate root canal treatment is done, these will be ultimate disappearance of granuloma and complete resolution of radiolucency to normal bone.
- *Periapical scar*—if granuloma has been subjected to repeated exacerbation due to periodic contact with the irritation from root canal it may become fibrosed. If root canal treatment is done successfully, remaining inflammation and granulomatous tissue will resolve leaving only fibrosed areas behind. Such entities are referred to periapical scar.
- *Osteomyelitis*—if the infection is not controlled there are chances of infection that may turn into osteomyelitis.

Radiographic Considerations

- *Lamina dura*—there are variations in lamina dura from person to person and region to region. Lamina dura is not continuous in case of periapical pathology.
- *Amount of bone destruction*—30 to 60% bone destruction required for radiographically detectable changes. Radiographic changes can not be detected without involvement of junctional and cortical bone.
- *Root resorption*—external root resorption is caused by orthodontic movement, inflammation or infection of bone and benign and malignant tumor.

Etiology of Major Facial Infection

Teeth

- *Decay (caries)* reaching the dental pulp—pulpitis, this in turn spreads to supporting bone, resulting in periapical abscess, which in turn may spread subperiosteally.
- *Periapical abscess* may occur in seemingly intact but devitalized teeth (trauma, cracks or decay under fillings).
- Periapical and periodontal abscess may form as a result of chronic gingivitis and supporting bone and soft tissue loss (*periodontal disease*).
- *Erupting teeth* (especially partially impacted lower third molars) can result in inflammation and infection of the

soft tissue flap preventing eruption (operculum) with resultant swelling, pus, etc. around the crown (pericoronitis).

Jaws

- *Cyst and tumors*—it can develop *cysts or tumors* that can range from odontogenic to either primary or secondary malignancy. Most are derived from the dental apparatus and although benign, can nevertheless continuously grow and become secondarily infected on breaching the surrounding bone.
- *Osteomyelitis*, although rare, can be the result of chronic infection, as mentioned before.
- *Osteoradionecrosis* occurs readily in irradiated jaws subjected to further trauma (such as extractions).
- *Bacterial infection*—these are tuberculosis, actinomycosis and syphilitic osteomyelitis.
- Most *jaw fractures* in the tooth bearing segments are defined to be compound to the oral cavity and can easily be infected by the oral microbes.
- *Extraction sites*, again are comparable to compound fractures and it is surprising that infection in such cases are relatively rare.

Major Salivary Glands

It may be the subject of either viral or bacterial infections, often superimposed on obstruction of ducts (stones, stricture, etc). Tumors, rarely, also becomes secondarily infected.

Paranasal Sinuses

It may be primarily infected, obstructed and result in facial swelling. It may become infected, secondary to infected teeth protruding into the maxillary sinus (upper premolars and molars often exhibit close proximity to sinus). Tumors or cysts may become infected. Fractures such as the orbital floor, are by definition compound to the “exterior” and may result in orbital cellulitis.

Acute and Chronic Infections of Oral Cavity

Periapical Abscess

An abscess is a localized collection of pus, surrounded by an area of inflamed tissue in which hyperemia and infiltration of leukocytes is marked. Abscess can be caused by trauma or chemical or mechanical irritation. Due to trauma, there is bacterial invasion of dead pulp.

Types

- *Acute or symptomatic*—it is localized collection of pus producing symptoms of severe pain in the tooth.
- *Chronic or asymptomatic*—it is a long standing, low grade infection of the periradicular tissues. Symptoms of chronic infection are less severe.

Pathogenesis

- *Microorganism*—streptococci viridans and staphylococci are frequently associated with abscess formation. They produce the enzyme called coagulase which causes fibrin deposition and thus helps in walling off the lesion. Coagulase promotes virulence by inhibiting phagocytosis.
- *Bacterial invasion*—bacterial invasion of pulp canal takes place. Canal contains large number of virulent bacteria that rapidly spread to periapical tissues, causing acute periodontitis, tender tooth and alveolar swelling.
- *Necrosis of periapical tissue*—when inflammatory response may extend into adjacent periapical alveolar bone, it will initiate necrosis of periapical tissue.
- *Abscess formation*—necroses tissue coalesces and enlarges, compressing the surrounding fibrous connective tissue. Thus an abscess is generated, which is a collection of pus surrounded by a wall of fibrous connective tissue.
- *Progress of abscess*—enlarging dentoalveolar abscess contains purulent material that is under pressure due to the production of pus. The purulent material travels along path of least resistance, until it reaches the surface, where due to limitation of periosteal layer, it temporally forms subperiosteal abscess. Eventually, it erodes through the periosteum and penetrates the soft tissue, again, following the path of least resistance. Path of least resistance is determined by the location of breakthrough in the bone and the anatomy of muscles and fascia plane in the area.
- *Sinus formation*—well circumscribed lesion may form sinus due to:
 - Inability of the body to completely contain or localized the causative organisms.
 - Increase in number of causative organisms.
 - Lowering of patient's general resistance.
 - Trauma or surgical intervention.

Clinical Features

Acute periapical abscess

- *Location*—it develops in the periapical region of the tooth as sequelae to necrosis of pulp (**Fig. 19-1**).
- *Symptoms*—pain is severe and of throbbing type. Periapical abscess may confine to osseous structures



Fig. 19-1: Periapical abscess showing swelling in palatal surface.

and during the early period of abscess formation, may cause excruciating pain without observable swelling. The patient may appear pale, irritable and weak from pain, loss of sleep as well as from absorption of septic products. Patient may have slight fever (99 to 100° F).

- *Signs*—patients experience sensitivity or pressure in the affected area. Ice relieves the pain and heat intensifies it. Aspiration yields yellowish pus.
- *Positive tenderness*—the tooth becomes more painful, appears elongated and mobile. In acute periapical infection, tooth is sensitive to percussion and movement.
- *Lymph node involvement*—there is also painful lymphadenopathy.
- *Swelling*—swelling is usually seen in adjacent tissues adjacent to the affected tooth (**Fig. 19-2**). The tissues at the surface of swelling appear to be taut and inflamed.



Fig. 19-2: Swelling involve palatal surface and extending into mid palatal region.

The surface of tissue become distended from the pressure of underlying pus and finally ruptures due to pressure and lack of resistance caused by continued liquefaction.

- **Maxillary involvement**—when the maxillary anterior teeth are involved, swelling of upper lip may extend to one or both eyelids. When the maxillary teeth are involved, the cheek may swell to an immense size, distorting the patient's face. When the maxillary posterior teeth are involved, there is possibility of maxillary sinus involvement (**Fig. 19-3**).
- **Mandibular involvement**—when the mandibular posterior teeth are involved, swelling of the cheek may extend to ear.



Fig. 19-3: Palatal abscess (Courtesy Dr Bhaskar Patle).

Chronic periapical abscess

- **History**—history of pain that started as dull ache and progressed to severe throbbing type. Sudden decrease in pain signals the formation of sinus.
- **Signs**—vitality test is negative with involved tooth.
- **Sinus formation**—sinus opening appears as a small ulcer (**Fig. 19-4**), rough and may bleed easily. Occasionally, after temporary emphysema, sinus heals and form slightly raised pale papule. As the pus accumulates, another sinus formation may take place eventually.
- **Location of draining fistulas or sinus**—these are also commonly associated with chronic alveolar abscess. Majority of sinus or fistula opens on labial and buccal aspect of alveolus, as apices of both maxillary and mandibular teeth are located nearer to the buccal than the lingual cortical plate. In maxilla, roots of lateral incisors and molars are close to palatal cortical plate, so sinus can appear there. Most root tips lie below the mylohyoid muscle, so pus drains into the submandibular space.
- **Parulis**—at the opening of sinus, there is mass of inflamed granulation tissue. This is called as Parulis.



Fig. 19-4: Intraoral sinus formation seen in case of chronic periapical infection.

- **Cutaneous sinus**—in some cases, dental abscess may drain into overlying skins and drain via cutaneous sinus (**Figs 19-5A and B**).



Figs 19-5A and B: Cutaneous sinus showing extraoral opening.

- *Lymph nodes*—the patients will demonstrate lymphadenopathy as well.
- *Turner hypoplasia*—it is present in case when infection from deciduous teeth spread to the successor teeth resulting in hypoplasia of tooth.

Radiographic Features

Acute periapical abscess

- *Periodontal ligament space widening*—swelling of periodontal ligament space force the tooth slightly from its socket, creating widening of periodontal ligament space (Fig. 19-6).

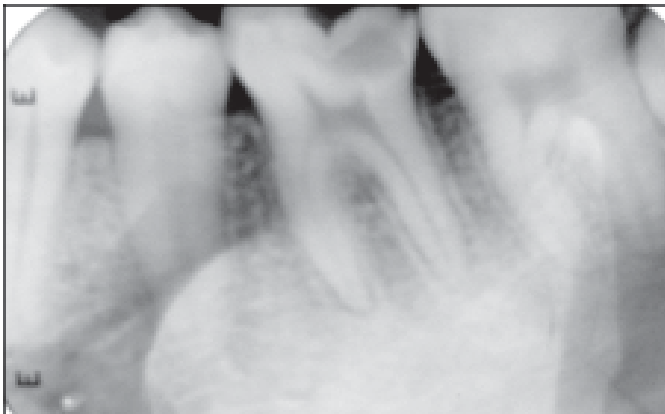


Fig. 19-6: PDL space widening seen in acute periapical infection.

- *Unsharpness at the tooth apex*—after some period, the first change seen is usually of slight unsharpness of some of the trabeculae at the tooth apex.
- *Cortex destruction of developing follicle*—if acute alveolar abscess of deciduous teeth occurs, the possibility of damage to the permanent successor can occur which is evident by rarefaction produced by destruction of the cortex of the follicle.

Chronic periapical abscess

- *Lamina dura*—there is loss of thickness and density of the apical portion of lamina dura of the affected tooth (Fig. 19-7).
- *Periapical rarefaction*—widespread area of diffuse demineralization of the periapical bone, of affected tooth becomes apparent (Fig. 19-8).
- *Margins*—margins vary from well defined with possible hyperostotic borders to poorly defined, in chronic cases. In some areas, margins fade gradually and imperceptibly into the normal bone, while in some there is abrupt demarcation between the normal bone and the radiolucent area.
- *Advanced cases*—after some period, the trabeculae which are rarefied are destroyed and the dark area become darker and larger, as more of the surrounding bone is taken into the diseased area (Figs 19-9A and B).



Fig. 19-7: Chronic periapical infection in upper first molar showing loss of lamina dura (Courtesy Dr Ashok L).

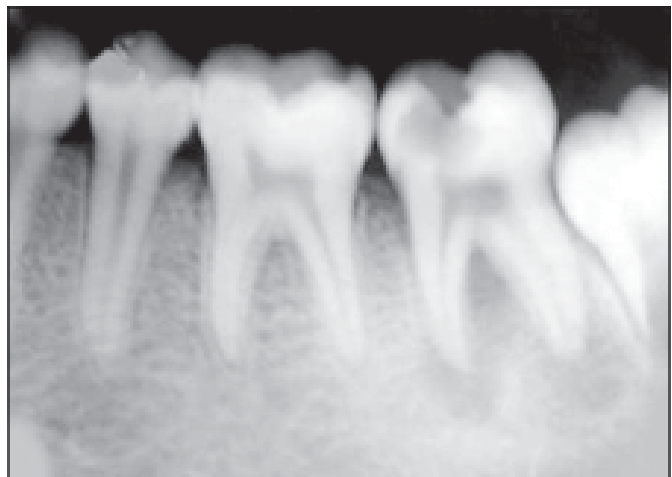
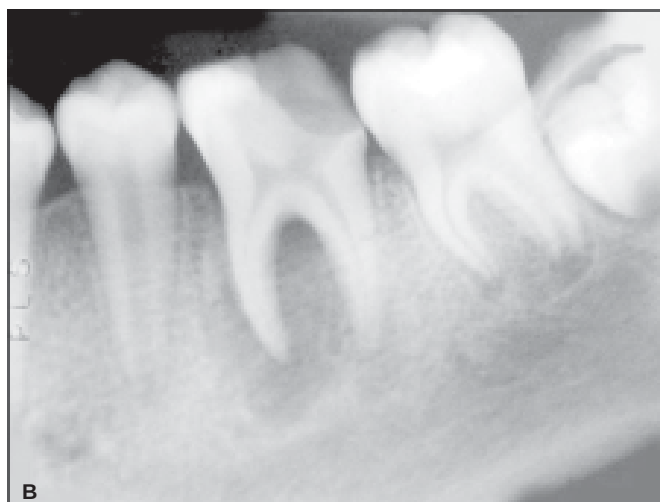
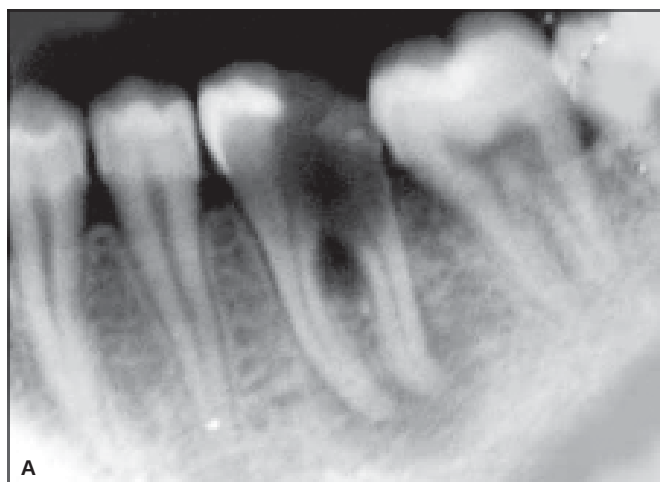


Fig. 19-8: Periapical rarefaction seen in chronic periapical infection.

- *Adjacent tooth*—in some cases, radiolucency may involve adjacent tooth and there may be loss of lamina dura of that tooth.
- *Side of root*—in some cases, osteitis can occur at the side of root rather than the apex. The reasons for this are that the infection may spread from adjacent tooth or perforation of the root (while filling the canal) or from an aberrant canal opening.
- *Maxillary upper posterior teeth*—in case of maxillary upper posterior teeth (as they are near to maxillary sinus), the radiographic appearance shows bone destruction at the apex of the tooth, along with loss of lamina dura and with destruction of a portion of the antral floor.
- *Root*—roots of the affected teeth may show resorption.

Diagnosis

- *Clinical diagnosis*—positive tenderness, carious tooth, sinus formation, non-vital teeth will give clue to the diagnosis.



Figs 19-9A and B: Loss of bone density seen in periapex of the tooth due to chronic periapical infection.

- **Radiological diagnosis**—PDL space widening, loss of lamina dura and periapical rarefaction will diagnosed this condition.
- **Laboratory diagnosis**—central region of necrosis contain dense accumulation of polymorphonuclear leukocytes, surrounded by inflamed connective tissue wall of varying thickness.

Differential Diagnosis

- **Periodontal abscess**—it is an accumulation of pus along the root surface of a tooth. It is associated with periodontal pocket with mild pain. It is usually associated with vital, rather than pulp less teeth.
- **Periapical osteofibrosis**—it is associated with vital tooth. There may be persistence of lamina dura in the periapical osteofibrosis, even in the presence of well marked bone destruction.
- **Foramina**—in it, lamina dura is intact. If foramens are exactly superimposed on the roots, then

another radiograph should be taken at different angulation.

- **Inferior dental canal**—it is frequently superimposed on the lower molars and if the canal is dark and apex is small, it is difficult to trace the lamina dura. In this case, magnifying glass should be used to trace the continuity of lamina dura.
- **A large normal marrow space**—it can be confused if superimposed on the apex of the root. Again, by using magnifying glass, lamina dura can be traced.

Management

- **Establish drainage immediately, if possible**—it may be done by opening the pulp chamber and passing file through the canal into the periapical region. Trephination opening through mucosa and bore to the abscess at apex. Through and through drain is placed in the abscess and irrigated with 1:1 mix of 3% H₂O₂ and normal saline solution.
- **Antibiotics** like Penicillin 500 mg, QID, for 5 days and analgesics should be given.
- **Endodontic treatment**—in 24 to 48 hours, it can be determined if the tooth can be treated endodontically or extraction is necessary. If there is need of retention of offending tooth, necrotic pulp should be opened and tooth should be treated endodontically.
- **Warm saline mouth rinse** often aid in localizing the infection and maintaining adequate drainage, before endodontic treatment or extraction.
- **Extraction of teeth**—the tooth which can not be restored should be extracted.

Acute Apical Periodontitis

When the inflammatory degradation products from infected pulp penetrate, apically periodontal ligament in sufficient amount, inflammation is initiated. In this condition there is no frank abscess formation present. This will help to differentiated it with acute periapical abscess.

Etiology

- **Trauma**—it may occur due to occlusal trauma caused by abnormal occlusal contact or by recently inserted restoration beyond the occlusal plane.
- **Wedging**—wedging of foreign object between the tooth such as toothpicks, food or a sliver of rubber dam left by teeth can cause inflammation.

Clinical Features

- **Location**—inflammation is restricted to periodontal ligament.

- *Symptoms*—pain is of throbbing type.
- *Signs*—tooth is non-sensitive to hot, cold, sweet or sour food. Due to apical edema, tooth is elevated in the socket. It is very tender to pressure and percussion.

Radiographic Features

- *PDL space widening*—widening of periodontal ligament space is caused by edema (Fig. 19-10).

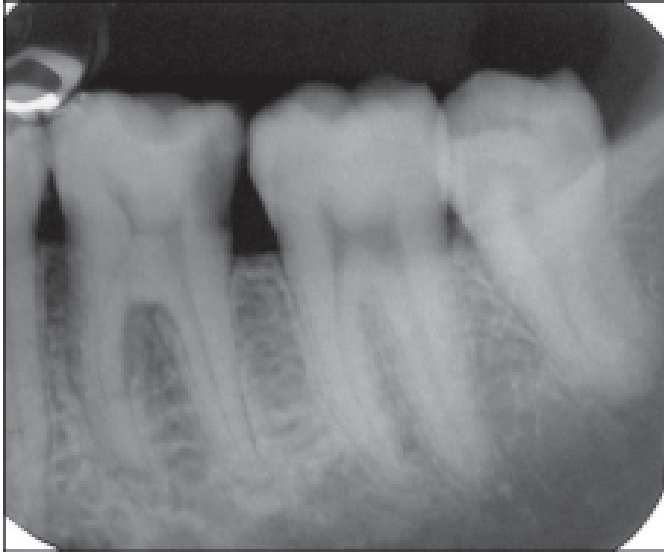


Fig. 19-10: Acute apical periodontitis seen with first molar (Courtesy Dr Ashok L).

Diagnosis

- *Clinical diagnosis*—tender tooth without abscess formation and secondary to trauma will give clue to diagnosis
- *Radiological diagnosis*—periodontal ligament (PDL) space widening is present.
- *Laboratory diagnosis*—an inflammatory reaction occurs in the apical periodontal ligament. The blood vessels are dilated. Polymorphonuclear leukocytes are present and an accumulation of serous exudate distends the periodontal ligament and extrudes the tooth slightly.

Management

- *Conservative management*—determine the cause and reliving the symptoms. When the acute phase is subsided, the tooth is treated by conservative means.

Acute Exacerbation of a Chronic Lesion

It is also called as '*phoenix abscess*.' It is an acute inflammatory reaction superimposed on an existing chronic lesion, such as on cyst or granuloma.

Etiology

The peri-radicular area may react to noxious stimuli form a diseased pulp with chronic peri-radicular disease. At times, because of an influx of necrotic product from a diseased pulp or because of bacteria and there toxins, this apparently dormant lesion may react and cause an acute inflammatory response.

Clinical Features

- *Symptoms*—patient complains of intense pain, local swelling and possibly associated cellulitis (Fig. 19-11).
- *Signs*—at the onset, tooth may be tender to touch.
- *Surface*—mucosa over the radicular area may be sensitive to palpation and may appear red and swollen.
- *History*—the patient has history of traumatic accident that turned the tooth dark after a period of time or of postoperative pain in a tooth that had subsided until the present episode of pain.
- *Vitality test*—lack of response to vitality test points to diagnose necrotic pulp.



Fig. 19-11: Local associated cellulitis occur in patient with phoenix abscess.

Radiological Features

Radiological features of this condition are same as that of chronic periapical infection (Fig. 19-12).

Diagnosis

- *Clinical and radiological diagnosis*—positive tenderness with radiological finding of chronic periapical infection will diagnose this condition.
- *Laboratory diagnosis*—area of liquefaction necrosis with disintegrating polymorphonuclear neutrophils and cellular debris. These are surrounded by infiltration of macrophages and some lymphocytes.



Fig. 19-12: Phoenix abscess showing same appearance as that of chronic periapical abscess.

Differential Diagnosis

- *Acute alveolar abscess*—vitality test; it will react to electrical pulp test and application of cold, as compared to acute exacerbation of chronic infection.

Management

- *Drainage*—either via the root canal or by incision, if there is localized swelling.
- *Antibiotics and anti-inflammatory drugs*—this should be given to the patient.

Pericoronal Abscess

It is also called as '*pericoronitis*'. It is the infection of soft tissues surrounding the crown of a partially erupted tooth.

Clinical Features

- *Location*—the most common type of pericoronal infection is found around the mandibular 3rd molar.
- *Symptoms*—pain, malaise is present. Pain may radiate to throat, ear or floor of mouth.
- *Sequelae of pericoronitis*—it may result in cellulitis and muscular trismus. There is also regional lymphadenopathy, submaxillary and pharyngeal abscess.
- *Signs*—operculum may get traumatized by opposing teeth during mastication (**Figs 19-13 and 19-14**). Edema, visible in both submandibular area and peritonsillar region. There is extreme tenderness on palpation of the abscess.
- *Pericoronal infection of infancy*—pericoronal infection of infancy is often associated with the supra-dental tissue, involving the superior portion of the follicle and the overlying mucoperiosteum, which may become



Fig. 19-13: Pericoronitis affecting partially erupted third molar.

inflamed. It ultimately develops into small fluctuant abscess. When this fluctuance is digitally ascertained, incision and drainage should be carried out, followed by warm saline rinses at frequent intervals.

- *Complication*—the involvement may become localized in the form of a pericoronal abscess. It may spread posteriorly into the oro-pharyngeal area and medially, to the base of the tongue. Peritonsillar abscess, cellulitis and Ludwig's angina are common.



Fig. 19-14: Pericoronitis showing inflammation of operculum around the third molar.

Radiographic Features

- *Appearance*—defect in bone on mesial or distal side which appears as the step like distortion of crypt wall distal to the crown (**Fig. 19-15**).
- *Lower 3rd molar area*—in cases of lower third molar, there is circumferential bone resorption around the tooth.

- *Sclerosing osteitis*—follicular crypt may show sclerosing osteitis. Generalized thickening of the wall of crypt indicates low-grade infection.
- *Semilunar shaped bone resorption*—mesially tipped impaction display semilunar shaped bone resorption mesial to the crown and in cases of distally placed impaction, it is distal to the crown.

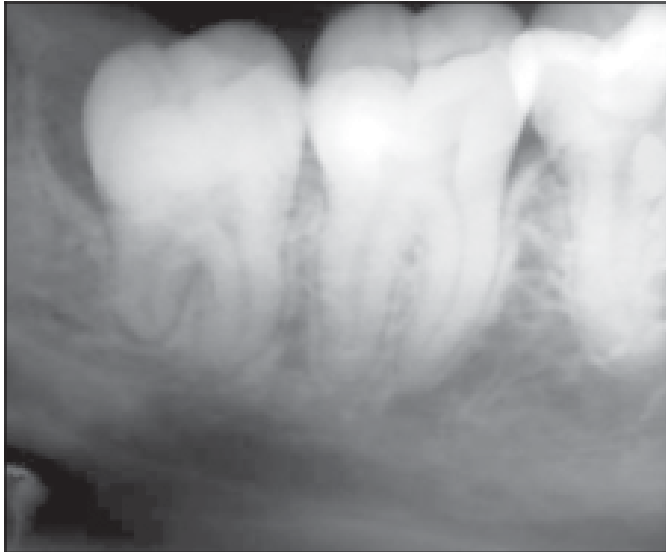


Fig. 19-15: Distal bone loss seen in association with 3rd molar suggestive of pericoronitis.

Diagnosis

- *Clinical diagnosis*—inflammation of pericoronal tissue around 3rd molar with pain and trismus will give clue to the diagnosis.
- *Radiological diagnosis*—there is semilunar shaped distal bone loss.

Management

- *Antibiotics*—immediately, antibiotics should be started. Most commonly use antibiotics are phenoxymethyl penicillin 250 mg four times daily. In pericoronitis due to ulceromembranous gingivitis metronidazole 200 gm three times daily for 7 days is given.
- *Drainage*—careful probing should be done around the 3rd molar, which permits entry into the expanded follicle and allows evacuation of pus and other septic material.
- *Extraction*—when the symptoms become sub-acute, the impacted 3rd molar should be extracted.
- *Operculectomy*—sometimes when the retention of 3rd molar is necessary, the inflamed tissue surrounding the occlusal portion of the tooth should be excised.
- *Extraction of maxillary 3rd molar*—maxillary 3rd molar can be a contributing factor to the pericoronal infection of the mandibular 3rd molar. In such cases, especially when the mandibular 3rd molar is fully erupted in

proper place, the maxillary 3rd molar should be extracted prior to the retention of the mandibular 3rd molar, considering the recurrent nature of the inflammatory episode.

Superficial Abscess

The yellow color is imparted by accumulation/pooling of pus in the enlarging abscess, covered by thin, stretched skin (**Fig. 19-16**). Fistula, if formed, drains the pus and no yellow color will be imparted.

It appears as raised sessile swelling with smooth, frequently reddened mucosa, over yellow pus. The most common presenting symptoms, is pain which may be slight or severe. Fluctuant swelling and when aspirated, yield pus. Surface may be ulcerated.



Fig. 19-16: Superficial infection of the facial tissue showing covered by stretched mucosa.

Cellulitis

It is also called '*Phlegmon*'. Occasionally, the infectious process progress out of the bone, despite the use of supportive therapy and the patient develops cellulites, either in vestibular region or extraorally. It is a potential complication of acute dental infection.

Cellulitis may be defined as a non-suppurative inflammation of the subcutaneous tissues extending along the connective tissue planes and across the intercellular spaces. It occur when abscess is not able to drain through surface of skin.

Bacteriology

- *Hemolytic streptococci*—the alpha hemolytic streptococci are the classic etiologic agent. They produce enzymes like streptokinase and hyaluronidase. These enzymes break down fibrin, connective tissue ground substance and cellular debris, thus facilitating rapid spread of bacterial invaders.



Fig. 19-17: Cellulitis showing diffuse involvement of buccal space.

Clinical Features

- *Symptoms*—there is widespread swelling, redness and pain without definite localization.
- *Signs*—there is presence of tenderness on palpation. Tissues are grossly edematous. There is marked induration, hence tissues are firm to hard on palpation. Tissues are often discolored; temperature is elevated with malaise and lethargy. Usually massive cellulitis (Fig. 19-17) will ultimately suppurate, particularly if bacteria are staphylococcal.
- *Evacuation of pus*—depending upon the location and proximity to anatomic structures that guide the progress, the pus may evacuate into nose, maxillary sinus, and oral vestibule, floor of mouth, infra-temporal fossa and into fascial spaces.
- *Maxillary infection*—infections arising in maxilla may perforate the outer cortical layer of bone, above the buccinator attachment and, cause swelling of upper half of face. If infection in mandible perforates the outer cortical palate, below the buccinator attachment, there is diffuse swelling of the lower half of face, which then spreads superiorly as well as cervically.
- *Eye*—if maxillary tooth is involved, there may be redness of eye (Fig. 19-18).

Diagnosis

- *Clinical diagnosis*—diffuse edematous swelling with positive tenderness will give clue to diagnosis.
- *Laboratory diagnosis*—ESR and white cell count are raised. Biopsy shows diffuse exudation of polymorphonuclear leukocytes and occasional lymphocytes, with considerable serous fluid and fibrin, causing separation of connective tissue or muscle fibers. It presents only a nonspecific picture of diffuse acute inflammation.



Fig. 19-18: Maxillary infection showing orbital involvement.

Management

- *Surgical incision and drainage*—it is performed when the presence of pus is diagnosed. In case of large cellulites, a superficial erythematous spot develops, which is pathognomonic of pus near the superficial surface. These superficial fluctuant areas can be incised and drained under local anesthesia. Usually ring block of peripheral skin, as a wheel is made for skin anesthesia. Knife is introduced in the most inferior portion of the fluctuant area. A small sinus forcep is introduced in the wound, opened in several directions and pus is drained. A rubber drain is placed in the deepest portion of the wound, so that just 12 cm lie above the source of the skin, where it is sutured. A large dressing is applied. When no superficial spot is present, fluctuance is more difficult to determine, particularly if deep pus is suspected.
- *Antibiotics*—heavy antibiotics should be given to the patient. Usually antibiotics in the cephalosporin family should be given.
- *Extraction*—extraction of the offending tooth should be carried out.

Ludwig's Angina

It is a condition which was first described by Ludwig in 1936. The word angina means sensation of choking or suffocation. It is the most commonly encountered neck space infection.

It is a rapidly swelling cellulitis of the sublingual and submaxillary spaces, often arising from infection of the

tooth roots (molars and pre-molars) that extend below the mylohyoid line of the mandible.

This condition may be defined as an overwhelming, rapidly spreading, septic cellulitis involving submandibular, submental and sublingual spaces bilaterally.

Etiology

- *Odontogenic infection*—it is usually an extension of odontogenic infection from mandibular molar teeth into the floor of mouth. The 2nd and 3rd molars are the teeth most commonly involved.
- *Trauma*—it may also be caused by oral soft tissue laceration and punctured wounds of the oral floor.
- *Sialadenitis*—submandibular gland sialadenitis and infected malignancy may be sometimes contributory to Ludwig's angina.
- *Calculi*—salivary calculi or from intravenous injection of the internal jugular vein, especially in drug abusers.
- *Osteomyelitis*—osteomyelitis in compound mandibular fracture.

Bacteriology

Streptococci are the most commonly reported organism from the culture. Other microorganisms are α -hemolytic streptococci, bacteroides, *Klebsiella*, *Fusiform bacilli* and *E. coli*.

Clinical Features

There are three typical appearances of Ludwig's angina.

First

- *Brawny induration*—it is characterized by brawny indurations. Tissues are board like and do not pit on pressure. No fluctuation is present.
- *Gangrenous tissue*—the tissues may become gangrenous and when cut, they have a peculiar lifeless appearance.
- *Limitation between infected and normal tissue*—a sharp limitation is present between the infected tissues and surrounding normal tissues.

Second

- *Spaces involve*—three facial spaces are involved bilaterally, i.e. submandibular, submental and sublingual. If the involvement is not bilateral, the infection is not considered a typical Ludwig's angina.

Third

- *Open mouth and respiratory obstruction*—the mouth is opened (**Fig. 19-19**) and the tongue is lifted upwards and backwards, so that it is pushed against the roof of the mouth and the posterior pharyngeal wall; when this occurs, acute respiratory obstruction is likely to occur.



Fig. 19-19: Typical open mouth appearance seen in Ludwig's angina patient.

Other Features

- *Signs*—swelling is firm, painful and diffused, with no sign of localization. Floor of mouth appears erythematous and edematous. Stiffness in tongue movement generally develops. The patient develops a toxic condition and speech becomes impaired. Larynx and glottis become edematous. As the disease continues the swelling starts involving the neck.
- *Symptoms*—the patient always has fever and there is considerable salivation, as the patient is unable to swallow. There are chills accompanied with fever. There is an inability to swallow and to eat. Respiratory distress is common. There is also neck pain, redness of neck, weakness, fatigue, excessive tiredness, confusion or other mental changes, difficult breathing and earache. There is an intense pain on tongue movement and the patient may be severely dehydrated, owing to inability to take anything by mouth. If the swelling has spread into the pterygoid region, then there is difficulty in opening the mouth.
- *Woody tongue*—due to involvement of sublingual space, there is elevation, posterior enlargement and protrusion of tongue. This is called as woody tongue.
- *Bull neck*—there is enlargement and tenderness of neck above the level of hyoid bone due to submandibular gland involvement. This is called as bull neck (**Fig. 19-20**).



Fig. 19-20: Bull neck seen in Ludwig's angina patient (Courtesy Dr Bhaskar Patle).

Fatal Complications

- *Respiratory obstruction*—the infection of Ludwig's angina tends to spread through the connective tissues which cover the small muscles of the larynx and between the muscles of the floor of mouth. The epiglottis and larynx become edematous along with the posterior aspect of the tongue. The tongue gets elevated and gets pressed upward and backward, causing pressure on the larynx. Therefore, dyspnea occurs in paroxysm. Ultimately, death occurs due to respiratory embarrassment.
- *Generalized septicemia*—this can occur due to spread of infection.
- *Erosion of the carotid artery*—late spread of infection to lateral pharyngeal space can also cause erosion of the carotid artery.
- *Cavernous sinus thrombosis* with subsequent meningitis may be sequelae of it.
- *Others*—mediastinum extension, pharyngomaxillary space extension, osteomyelitis and airway obstruction.

Diagnosis

- *Clinical diagnosis*—open mouth appearance, woody tongue, bull neck and involvement of three spaces will diagnose this condition.
- *Laboratory diagnosis*—a moderate leukocytosis is found. Fusiform bacilli and spiral forms, various staphylococci, diphtheria and many other microorganisms have been cultured on different occasions.

Management

- *Intense and prolonged antibiotic therapy*—high dose penicillin is to be administered IM or IV, in high doses, because it is the empirical antibiotic of choice and the oral flora, including most of anaerobes, are sensitive to

it. Recently, combination of gentamicin and cloxacillin has been proved to be successful. In case of person who is resistant to penicillin, clindamycin, aminoglycoside and chloramphenicol can be given.

- *Establishment and maintenance of an adequate airway are the essentials of therapy*—tracheostomy is a routine procedure, but it is often difficult to perform in the late stages. Attempt at blind intubations is often time consuming. Recently, successful intubations with fiberoptic laryngoscope have been introduced as a worthwhile technique in the therapy of Ludwig's angina. In the late stage, cricothyroidotomy may be performed as an emergency procedure. Nowadays, administration of corticosteroids allows more rapid penetration of antibiotics and protection of airway. Most commonly used corticosteroid is intravenous dexamethasone should be given.
- *Incision and drainage*—it is done to release the tissue tension. A horizontal incision, midway between the chin and hyoid bone, is a classic approach to surgical drainage of Ludwig's angina. It may be carried out under local anesthesia.
- *Supportive therapy*—parenteral hydration, high protein diet and vitamin supplements.
- *Extraction of offending tooth*—extraction of offending tooth should be carried out.
- *Prevention*—regular visits to the dentist and prompt treatment of oral/dental infections can prevent the conditions that increase the risk of developing Ludwig's angina.

Impetigo

It is also called as 'non-bullous impetigo' or 'impetigo vulgaris'. It is acute superficial, purulent infection of the skin. It is caused by *Streptococcus pyogenes*.

Predisposing Factors

- *Poor oral hygiene*—this will lead to growth of organisms.
- *Living condition*—crowded living conditions and hot and humid climate is also responsible for impetigo.
- *Pre-existing disease*—pre-existing eczema and scabies and previous trauma and insect bite may also lead to impetigo.
- *Reduced resistance*—reduced resistance due to preceding influenza or herpes simplex infection.

Clinical Features

- *Age*—the disease is mainly seen in pre-school children and young adults.
- *Locations*—the face (angle of mouth, lips and nose) is the most common location.

- **Onset**—the lesions frequently begin as red, itchy spots.
- **Stuck on appearance**—the close set, round or oval, flat, pustular vesicles with a characteristic stuck on appearance subsequently develop.
- **Signs**—the thin roofed vesicle then burst to form golden yellow crust and some of the lesions may become confluent (**Fig. 19-21**).



Fig. 19-21: Impetigo showing rupture vesicle with some of them become confluent (Courtesy Dr Pincha).

- **Others**—the surrounding skin is erythematous with lymphadenopathy, which becomes tender.
- **Clinical pattern**—two clinical patterns are present. First is fragile vesicle which quickly ruptures (**Fig. 19-21**) and are replaced by thick adherent amber crusts and second long lasting bullous lesion.

Diagnosis

- **Clinical diagnosis**—red itchy sopt with typical stuck on appearance is characteristic of impetigo.

Management

- **Antibiotics**—single injection of long acting penicillin and 3-4 gm of oral penicillin V, daily, for 10 days. Alternate antibiotics which can be used are clindamycin, cephalexin and dicloxacillin.
- **Topical mupirocin**—it is effective treatment of choice.

Erysipelas

It is an acute, superficially spreading infection of the dermis, usually of the face, with a well demarcated, slightly indurated erythema and progressive lymphangitis. It is caused by Beta hemolytic streptococci. It is sometime also called as 'saint Anthony's fire' (as the wall of Saint Anthony's fire is that of same color of erysipelas).

Etiopathogenesis

- **Entry of microorganism**—the microorganisms are thought to enter the tissues through a small break in the mucosa or the skin, such as fissure, abrasion, erosion or excoriation.
- **Post-surgical erysipelas**—post-surgical erysipelas is caused by transmission of streptococci from the nose, throat, or hands of the patient or from, the attendant or visitors.
- **Predisposing factors**—nephrotic edema, lymphedema, dysgammaglobulinemia, malnutrition and alcoholism are known predisposing factors.

Clinical Features

- **Age and sex**—the newborn and infants are highly susceptible, but elderly are also affected. It is more common in women in 5th decade and in men, in 7th decade.
- **Location**—the most common location is on abdomen, face, scalp and legs.
- **Incubation period**—the incubation time it thought to be from few hours to several days.
- **Prodromal phase**—after short prodromal phase, characterized by malaise, vomiting, headache, pyrexia and chills. Erysipelas begins abruptly with a local sensation of burning and itching.
- **Symptoms**—the general condition of the patient worsens with toxemia, high fever, insomnia and restlessness.
- **Signs**—a small area of the skin then becomes intensely red and swollen (**Fig. 19-22**). Subsequently, bright red plaques develop and spread rapidly. Blisters, which break down to form large ulceration, may develop on the red lesion.
- **Margins**—the lesion has an elevated, edematous, sharp border and an irregular outline.
- **Lymph nodes**—regional lymph nodes become enlarged and tender.



Fig. 19-22: Red erythematous skin seen on the right cheek (Courtesy Dr Sanjay Pincha).

Oral Manifestations

- *Location*—it is uncommon in oral, pharyngeal and nasal mucosa. Erysipelas which develops in the oral, pharyngeal or upper respiratory tract mucosa result in the same constitutional reaction, as described for skin lesion.
- *Symptoms*—there is severe local pain, redness and swelling.
- *Signs*—edema may involve the tongue, uvula, epiglottis and may lead to serious consequences.
- *Lymph nodes*—the submandibular lymph nodes become markedly tender and swollen.

Diagnosis

- *Clinical diagnosis*—high fever with red and swollen skin with lymphadenopathy will aid in diagnosis.

Management

- *Antibiotics*—penicillin is the drug of choice and if sensitive to penicillin, erythromycin can be given.

Pyostomatitis Vegetans

It is an uncommon inflammatory disease of the oral cavity. It is unusual oral expression of inflammatory bowel disease. The name is given as there is a similarity between it and the skin lesions in a dermatologic disease known as pyodermitis vegetans. It may occur due to intestinal disturbances.

Clinical Features

- *Age and sex*—it occurs in any age with no sexual predilection.
- *Site*—this lesion may occur in any area of oral cavity, except in tongue. It is multiple in numbers.
- *Appearance*—oral lesions consists of large number of broad base papillary projections, tiny abscess or vegetations developing in areas of intense erythema.
- *Symptoms*—it painless but in some cases, oral discomfort may be present.
- *Signs*—these small projections are red or pink in color, but on careful examination may show tiny pustules beneath the epithelium, which liberate a purulent material when ruptured. Buccal and labial mucosal lesions have many folds and papillary projections may develop on these folds.
- *Snail track ulceration*—after vesicle is rupture ulceration is present which may coalesce into larger areas of necrosis.
- *Cobblestone appearance*—buccal mucosa show 'cobblestone' appearance, while vestibular lesions

appeared as folds and ulcers, the lips are diffusely swollen and indurated, gingival and alveolar mucosal lesions are granular with erythematous swelling and palatal lesions appear as multiple aphthous ulcers.

Diagnosis

- *Clinical diagnosis*—snail track ulceration with cobblestone appearance may give clue to the diagnosis.
- *Laboratory diagnosis*—biopsy shows papillary projections with tiny areas of focal necrosis and micro abscess formation, either intraepithelial or sub-epithelial.

Management

- *Sulfa drugs*—It is found that the oral lesions are regressed when the intestinal disturbance is brought under control. Medical management with sulfasalazine can be helpful in clearing the lesion.
- *Corticosteroids*—systemic steroids like prednisone is given in tapering doses. It is not specific.
- *Other therapy*—other therapy like fluocinonide gel and colectomy can be carried out.

Maxillary Sinusitis

It is described in chapter of paranasal sinus.

Chronic Hyperplastic Pulpitis

It is also called as 'pulp polyp' or 'pulpitis aperta'. It is essentially an excessive, exuberant proliferation of chronically inflamed dental pulp tissue.

Etiology

- *Slow carious exposure*—slow progressive carious exposure of the pulp is the cause.
- *Chronic low grade infection*—for the development of a hyperplastic pulp, a large open cavity, a young resistant pulp and a chronic low grade stimulus is necessary.
- *Mechanical irritation*—mechanical irritation from chewing and bacterial infection often provides the stimuli.

Clinical Features

- *Site*—teeth most commonly involved are deciduous molars and first permanent molars as they have an excellent blood supply because of a large root opening, and this coupled with high tissue resistance and reactivity in young person's accounts for unusual proliferative properties of the pulp tissue.
- *Age*—it is seen only in teeth of children and young adults.
- *Symptoms*—it is asymptomatic and there may be feeling of pressure when masticator forces are applied.

- **Appearance**—polypoid tissue appears as a fleshy, reddish pulpal mass filling most of the pulp chamber (**Fig. 19-23**) or cavity or even extending beyond the confines of the tooth.
- **Signs**—some times, mass if large enough interferes with comfortable closure of teeth. Polypoid tissue is less sensitive than normal pulp tissue and more sensitive than the gingival tissue. This tissue bleeds easily because of rich network of blood vessels.
- **Thermal testing**—the tooth may respond feebly or not at all to the thermal test.



Fig. 19-23: Chronic hyperplastic pulpitis presenting as reddish mass in third molar region (Courtesy Dr Tapasya).

Radiographic Features

It will show a large open cavity with direct access to the pulp chamber.

Diagnosis

- **Clinical diagnosis**—Fleshy pulpal mass in pulp chamber with negative thermal test will aid in diagnosis.

Differential Diagnosis

- **Proliferating gingivitis**—one should raise and trace the stalk of the tissue back to its origin, i.e. the pulp chamber.

Management

- **Pulp extirpation**—elimination of polypoid tissue, followed by extirpation of the pulp. After removing the hyperplastic tissue, bleeding can be controlled by pressure.
- **Extraction**—extraction of tooth can also be done.

Periapical Granuloma

It is the most common type of pathologic radiolucency encountered in dentistry. It is a growth of granulation

tissue continuous with the periodontal ligament resulting from the death of the pulp and diffusion of bacteria and bacterial toxins from the root canals into the surrounding periradicular tissues through the apical and lateral foramina.

It is the result of a successful attempt by the periapical tissues to neutralize and confine the irritating toxic product that is escaping from the root canal. But continuous discharge into the periapical tissues induces a vascular inflammatory response.

Etiopathogenesis

- **Prolonged irritation**—it occurs as a response to intense and prolonged irritation from infected root canals producing extension of chronic apical periodontitis beyond the periodontal ligament.
- **Release of inflammatory mediators**—insult from diseased pulp represents broad spectrum of inflammatory mediators like prostaglandins, kinin and endotoxins. Elevated level of IgG in pulpoperiapical lesion.
- **Replacement of bone by granulation tissue**—the expanding inflammation and increased vascular pressure result in abscess formation and resorption of the bone in the affected area, which in period of time is replaced by granulation tissue.

Clinical Features

- **Symptoms**—mild pain can be occasionally experienced while biting or chewing on solid foods.
- **Signs**—tooth may be darker in color, because of the blood pigments that diffuse into the dentinal tubules. There is, seldom, swelling or expansion of the overlying cortical bone. Tooth may feel to be slightly elongated in the socket.
- **Vitality test**—the tooth is non-vital, i.e. it does not respond to thermal and electric pulp test.
- **Sensitivity**—sensitivity occurs due to hyperemia, edema and inflammation of the apical periodontal ligament.

Radiographic Features

- **Lamina dura**—periapical area is radiolucent with loss of lamina dura (**Fig. 19-24**).
- **Size**—radiolucency is less than 1.5 cm in diameter (**Fig. 19-25**).
- **Margins**—there may or may not be hyperostotic borders. It may or may not have well defined borders.
- **Teeth**—involved tooth may show a deep restoration, extensive caries, fracture or a narrow pulp canal with non-vital pulp.
- **Multiple periapical granuloma**—in some cases, multiple periapical granuloma may be present (**Fig. 19-26**).

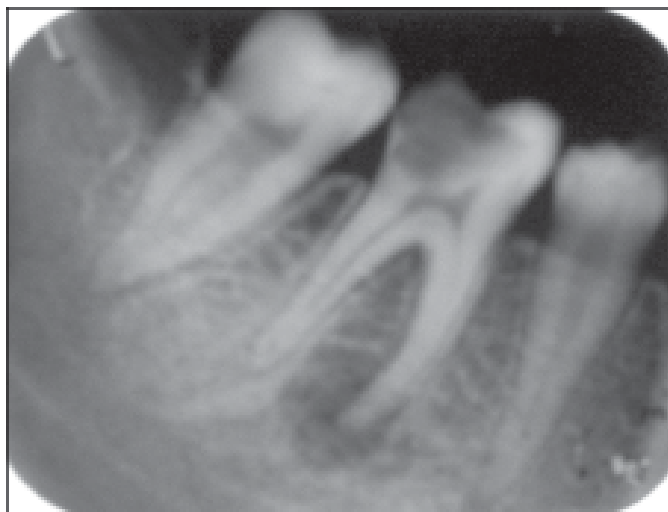


Fig. 19-24: Periapical granuloma presented as radiolucency in the periapex of tooth.

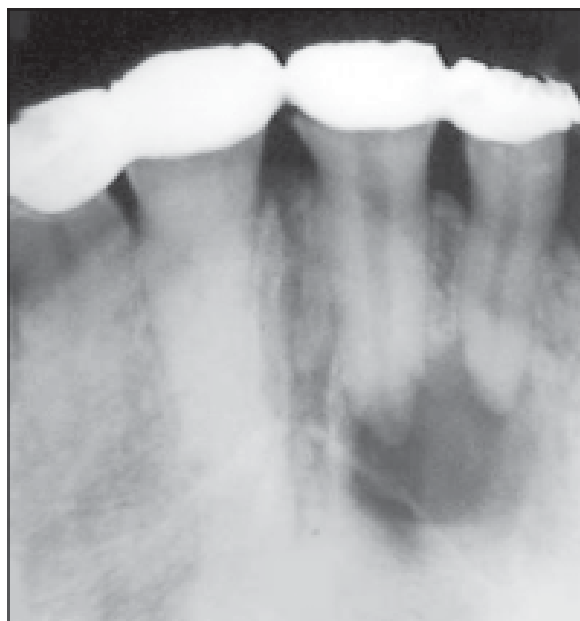


Fig. 19-25: Periapical granuloma presenting as radiolucency in relation with two teeth.



Fig. 19-26: Multiple periapical granulomas seen at the apex of the tooth.

Diagnosis

- *Clinical diagnosis*—it is not possible to make clinical diagnosis on the basis of signs and symptoms.
- *Radiological diagnosis*—well defined radiolucency less than 1.5 cm will go in favor of periapical granuloma.
- *Laboratory diagnosis*—biopsy shows proliferating endothelial cells capillaries, young fibroblasts minimum amount of collagen and occasionally, nests of odontogenic epithelium, Russell's bodies, foam cells and cholesterol clefts.

Differential Diagnosis

- *Osteolytic stage of Cementoma*—in case dental granuloma, tooth is non-vital.
- *Radicular cyst*—it is described in differential diagnosis of radicular cyst.
- *Surgical defect or periapical scar*—tooth shows root canals filling.

Management

- *Root canal therapy*—root canal therapy is treatment of choice.
- *Extraction of teeth*—if the tooth is unrestorable then extraction of the involved tooth should be carried out.
- *Curettage*—curettage of apical tissue can be carried out.
- *Causes of failure of treatment*—lesion may heal due to inadequate endodontic, vertical foreign material, associated periodontal disease, and cysts formation.

Periapical Scar

It is a possible end point of healing. It is composed of dense fibrous tissue and is situated at the periapex of pulp less tooth, in which usually, the roots canal have been successfully filled.

Formation of Scar

- *Irritant substance*—confined in the periapical area, which leads to accumulation of chronic inflammatory cells.
- *Granuloma formation*—young fibroblasts, endothelial cells and capillaries proliferate, which lead to granuloma formation.
- *Scar formation*—after endodontic treatment, the granuloma resolves, but in some cases, granulation tissue gets slowly organized with the production of more and more collagen fibers, which in turn leads to scar formation.

Clinical and Radiological Features

- *Location*—it is more common in anterior region of maxilla.
- *Tooth*—tooth is non-vital and the patient is asymptomatic.

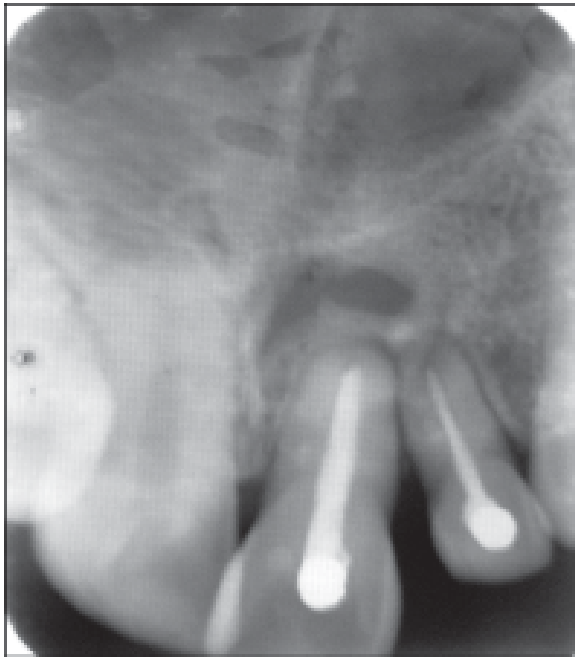


Fig. 19-27: Periapical scar seen as radiolucency at the apex of central incisor (Courtesy Dr Mody).

- **Radiolucency**—well circumscribed radiolucency that is more or less round and is smaller than granuloma and cyst (**Fig. 19-27**).
- **Size**—scar is constant in size throughout the period.

Diagnosis

- **Clinical diagnosis**—it is not possible to make clinical diagnosis.
- **Radiological diagnosis**—radiolucency at the apex of endodontically treated tooth.
- **Laboratory diagnosis**—biopsy shows spindle shaped fibroblast scattered throughout the dense collagen bundles, which show advanced degree of hyalinization.

Management

There is no treatment necessary for this disease

Osteomyelitis

Osteomyelitis is the inflammation of the bone marrow that produces clinically apparent pus and secondarily affects the calcified components. It is infection of the bone that involves all the three components viz. periosteum, cortex, and marrow.

It may be defined as an inflammatory condition of the bone that begins as an infection of medullary cavity and the haversian system and extends to involve the periosteum of the affected area.

Predisposing Factors

Certain predisposing factors play an important role in the onset and severity of the osteomyelitis, in addition to the virulence of microorganism.

- **Conditions affecting host resistance**—condition like diabetes mellitus, tuberculosis, severe anemia, leukemia, agranulocytosis, acute illness such as influenza, scarlet fever, typhoid and exanthematous fever, sickle cell anemia, malnutrition, and chronic alcoholism will affect host resistance. This will predispose for osteomyelitis occurrence.
- **Condition affecting jaw vascularity**—metastasis from remote area of infection such as another bony site, skin and kidneys, radiation, osteoporosis, osteopetrosis, fibrous dysplasia, bone malignancy, and peripheral vascular disease will affects jaw vascularity.

Etiology

- **Odontogenic infections** which can be periapical or periodontal infection, pericoronal infection and infection from infected dental cyst.
- **Compound fractures of the jaws**—generally, these fractures are compound through the tooth socket into the mouth and rarely, to the skin.
- **Traumatic injury**—local traumatic injury of the gingiva leads to periostitis, in patients with low resistance to infection and later to osteomyelitis.
- **Middle ear infection and respiratory infection**—via hematogenous route, either from middle ear infection or from infection of the upper respiratory tract.
- **Furunculosis of chin**—furunculosis of chin, i.e. spread through lymphatic channel via infected lymph nodes.
- **Peritonsillar abscess**—peritonsillar abscess has also been reported to cause osteomyelitis of the ramus of mandible.

Pathogenesis

- **Compromised blood supply** is a critical factor in the establishment of osteomyelitis.
- **The virulent microorganisms** get entry in the medullary cavity via many routes like odontogenic infections, compound fractures, periostitis, hematogenous route and lymphatic channel.
- **Inflammatory reaction**—these microorganisms cause intense inflammatory reaction within the marrow of the bone. Pain is a feature of this stage.
- **Localization of infection**—most of the odontogenic infections, like periapical and periodontal infections, are localized by pyogenic membrane or soft tissue abscess walls.
- **Disorganization of clot**—however, disorganization of this pyogenic membrane occurs by virulent microorganism

or by chronic movement of the unreduced fractures of jaws.

- **Mechanical trauma**—mechanical trauma, due to chronic movement of unreduced fractures, burnishes the bone and causes ischemia, thereby introducing the microorganisms deep into the underlying tissues.
- **Accumulation of pus**—when this protective barrier breaks, the pus accumulated in the medullary cavity gives rise to increased intra-medullary pressure, which results in compression of vasculature, vascular collapse, venous stasis and ischemia.
- **Elevation of periosteum**—pus travels through the Haversian and Volkmann's canals and accumulates beneath the periosteum, elevating it from the cortex, thereby further reducing the blood supply.
- **Necrosis of bone**—the reduced blood supply leads to slow necrosis of the bone.
- **Penetration of periosteum**—if the pus continues to accumulate, the periosteum is penetrated and mucosal and cutaneous fistulae develop and thereby discharging the purulent pus.
- **After therapy**—as the therapy begins to be effective and the host resistance increases, the process become chronic. Inflammation regresses, granulation tissue forms and new blood vessels are formed which cause lysis of bone; thus causing fragments of necrotic bone from the viable bone.
- **Involucrum**—small sections of necrotic bone may be completely lysed, while large one get localized and get separated from the shell of the new bone by a bed of granulation tissue. This dead bone surrounded by viable bone is called involucrum.
- **Sequestra**—small pieces of necrotic bone are called as sequestra, which are avascular and which harbor microorganisms. These sequestra need to be removed, otherwise they continue to be chronically infected and infect the surrounding granulation tissue and cause further sequestration, which weakens the bone and may cause pathologic fracture.
- **Cloacae**—an involucrum contains one or more holes on the surface which lead into channels, which can be traced to end in the depth of the bone at the site of an area of bone destruction around the sequestrum. These orifices are termed 'cloacae'. Pus finds its way from the depth of the bone to the surface, through the cloacae. The presence of cloacae indicates that there is a dead bone or a foreign body at the deep end.
- **Systemic spread of infection**—besides the pathogenic activity of the virulent microorganisms, these microorganisms precipitate thrombi formation by virtue of their destructive lysosomal packages. The coagulum provides the host medium for further pathogenic proliferation as well as an isolating barrier from the host immune response. It also allows systemic spread of infection.
- **Necrosis of bone**—the necrosis of bone is brought about by thrombosis of the vessels or compression of the vasculature. The necrosis of bone, with superadded infection forms a base line pathogenesis of the osteomyelitis. Compression of neurovascular bundles can result in osteomyelitis mediated anesthesia. The possibility of pre-existing predisposing factors can further complicate this phenomenon.
- **Remodeling**—after complete removal of sequestra and resolution of disease process, the jaw undergoes complete remodeling.

Microbiology of Osteomyelitis

- **Microorganism**—microorganism responsible for osteomyelitis are *Staphylococcus aureus*, *Staphylococcus albus*, hemolytic streptococci, gram-negative organisms like *Klebsiella*, *Pseudomonas*, *Protease*, *E. coli*, anaerobic microorganisms like pepto-streptococci, bacteroides and fusobacteria. Some specific forms like *Mycobacterium tuberculosis*, *Treponema palladium* and *A. israeli*.
- Following parameters should be used for recognition of pure anaerobic or mixed anaerobic infection:
 - Presence of foul smelling exudate.
 - Sloughing of necrotic tissues or gas in the necrotic tissue.
 - Gram stain revealing multiple organisms of different morphological character.
 - Presence of sequestra.

Classification

- **According to anatomic location of infectious process**
 - Intramedullary
 - Subperiosteal
 - Periosteal
- **According to duration and severity**
 - **Acute**—it occurs initial infection with the microorganism
 - **Chronic**—it can be primary or secondary
 - **Primary**—virulence of the microorganism is low and the host resistance is high. This type is not preceded by an episode of acute symptoms.
 - **Secondary**—it is secondary to incompletely treated acute osteomyelitis.
- **Depending upon the presence or absence of suppuration**
 - **Suppurative:**
 - Acute suppurative osteomyelitis
 - Chronic suppurative osteomyelitis
 - Infantile osteomyelitis
 - **Nonsuppurative:**
 - Chronic nonsuppurative
 - Focal sclerosing
 - Diffuse sclerosing

- *Radiation osteomyelitis*
- *Garre's sclerosing osteomyelitis*
- *Osteomyelitis due to specific infection:*
 - Actinomycosis
 - Tuberculosis
 - Syphilis.

Clinical Staging of Osteomyelitis

- *Initial stage*—spontaneous pain (localized).
- *Acute stage (suppurative stage)*—in this stage, there is severe pain, soreness and looseness of the involved teeth.
 - *Early acute stage*—in reference of to the involved tooth, progressive sensitivity of the adjacent teeth to percussion and pain in the involved side of jaw.
 - *Late acute stage*—paresthesia or anesthesia of the lip region supplied by the mental nerve. Other systemic symptoms can occur.
- *Osteonecrotic stage*—diminished spontaneous pain, abscess formation and pus discharge (**Fig. 19-28**).
- *Sequestrum stage*—lack of symptoms sequestrum formation visible on the radiograph.



Fig. 19-28: Osteomyelitis in osteonecrotic stage showing extraoral discharging sinus.

Occurrence

- *Sex*—it is more common in men, than women.
- *Sites*—it occurs in mandible in premolar area because:
 - The cortical plate of the bone in time mandible is dense. It takes longer time for sinus formation and release of pits and hence, the infection gets directs into spongiosa and spreads.
 - Removal of posterior mandibular teeth is attended by more damage to the bone.
 - Mandible is less vascular than maxilla.
 - Thin cortical plates and relative paucity of medullary tissue in the maxilla precludes confinement of infection within the bone and permits dissipation of edema and pus into the soft tissues and paranasal sinuses.
- *Infantile osteomyelitis* is more common in maxilla than mandible, as it spreads by hematogenous route and maxilla has more blood supply than mandible.

Clinical Features

Acute

- *Initial symptoms*—it has rapid onset and course. Patient complaint of severe pain, paresthesia or anesthesia of the mental nerve.
- *Initial signs*—at this stage, the process is truly intra-medullary, therefore swelling is absent, teeth are not mobile and fistulae are not present.
- *Late symptoms*—there is deep intense pain, anorexia, malaise, fever, and regional lymphadenopathy. Patient also complain of soreness of involved teeth which become loose within 10 to 14 days. There is also fetid oral odor.
- *Late signs*—pus exudates around the gingival sulcus or through mucosal and cutaneous fistula. There is firm cellulitis of cheek and abscess formation with localized warmth and tenderness on palpation. The patient feels toxic and dehydrated.

Chronic

- *Onset*—it has insidious onset with slight pain, slow increase in jaw size and a gradual development of sequestra without fistula.
- *Symptoms*—it is painless unless there is an acute or sub-acute exacerbation.
- *Necrotic bone*—in some cases, necrotic bone may be visible inside the oral cavity (**Figs 19-29A and B**).
- *Sinus*—intraorally and extraorally sinus develops (**Figs 19-30A and B**) intermittently and drains small amount of pus and then gradually heals. Sinus extends from medullary bone, through cortical plate, to mucous membrane or skin. Sinus may be at a considerable distance from the offending infection.
- *Signs*—local tenderness and swelling develop over the bone in the area of abscess (**Fig. 19-31**).
- *Lymph nodes*—regional lymphadenopathy is present.

Radiographic Features

Acute

- *Radiodensity*—about 10 days after acute infection, the density of trabeculae will be decreased, with blurred and fuzzy. For the radiographs to reveal any changes, there must be a loss of from 30 to 60% in the calcium content.
- *Trabecular pattern*—the earliest radiographic change is that trabeculae in the involved area are thin, of poor density and slightly unsharp or blurred. The trabeculae soon lose their continuity as well as the little density present. Individual trabeculae become fuzzy and indistinct.



Figs 19-29A and B: Necrotic bone seen in osteomyelitis.

- *Multiple radiolucency*—subsequently, multiple radiolucencies appear which become apparent on radiograph. These are enlarged trabeculae spaces caused by foci of necrosis and frank bone destruction.
- *Saucer shaped destruction*—in some cases, there is a saucer shaped area of destruction with irregular margins and containing teeth, with variable amount of supporting bone.
- *Periosteal reaction*—it can either stimulate bone resorption or bone formation. Inflammatory exudate can lift the periosteum and stimulates bone formation. Radiologically, it appears as a thin faint, radiopaque line, adjacent to and almost parallel or slightly convex to the surface of the bone.
- *Lamina dura*—there is loss of continuity of lamina dura, which is seen in more than one tooth.
- *Technetium bone scan followed by gallium citrate scan*—it will help to confirm the diagnosis. With inflammatory lesion, a positive result on the Tc^{99} scan indicates increased bone activity and a positive result on gallium scan in the same location indicates an inflammatory reaction.



Figs 19-30 A and B: Extraoral discharging sinus seen in osteomyelitis (Courtesy Dr Bande).



Fig. 19-31: Swelling seen in alveolar bone in case of osteomyelitis of mandible (Courtesy Dr Bande).

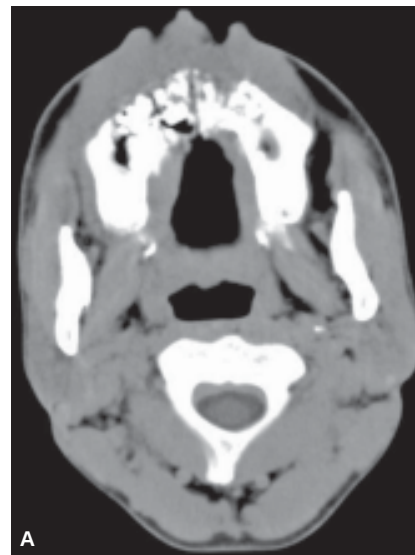
- *Osteomyelitis in children*—it shows paucity or absence of trabeculae in the tooth bearing area. In some cases, there may be loss of density on a part or whole of the cortical layer of one or more tooth follicles. Follicular cortex may become fragmented or lost over the variable area. Following destruction of the wall of the tooth follicle, it is common for the teeth to show evidence of moving.

Chronic

- *Radiodensity*—single or multiple radiolucencies of variable sizes are seen.
- *Margins*—irregular outline and poorly defined borders.
- *'Moth eaten appearances'* is seen as the radiolucent areas enlarge and become irregular in outline and get separated by islands of normal appearing bone. This is due to the enlargement of medullary spaces and widening of Volkmann's canals, secondary to lysis of bone and replacement with granulation tissue.
- *Sequestra*—segments of necrotic bone become detached; irregular calcified areas separate from the remaining bone and become distinguishable as sequestra. Sequestra are more dense and better defined (**Figs 19-32A and B**) due to following reasons:
 - Sclerosis that was induced before the bone became necrotic.
- Dead bone has affinity for calcium. Hence, it absorbs calcium.
- Inflammatory reaction is the probable stimulation for demineralization of the vital bone surrounding the sequestra. This enhances the contrast.
- *Teeth*—the roots of the teeth may undergo external resorption and the lamina dura may become less apparent as it blends with surrounding granular sclerotic bone.
- *Fistula tract*—fistula tracts may appear on the radiograph as radiolucent bands transversing the body of the jaw and penetrating the cortical plates.
- *Joint involvement*—in patients with extensive chronic osteomyelitis, the disease may spread to mandibular condyle and joint, resulting in septic arthritis.
- *Computed tomography*—computed tomography is more useful in revealing the internal structure and sequestra more readily than conventional radiography (**Figs 19-33 and 19-34**).



Figs 19-32A and B: Sequestration seen as horizontal radiopaque structure in the mandibular bone.



Figs 19-33A and B: Computed tomography showing destruction of mandibular cortex.

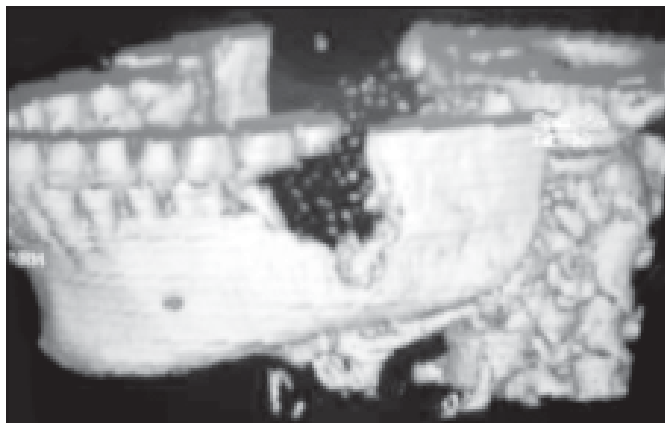


Fig. 19-34: Three dimensional CT of osteomyelitis.

- *Pathological fracture*—in some cases osteomyelitis can lead to pathological fracture (Fig. 19-35).

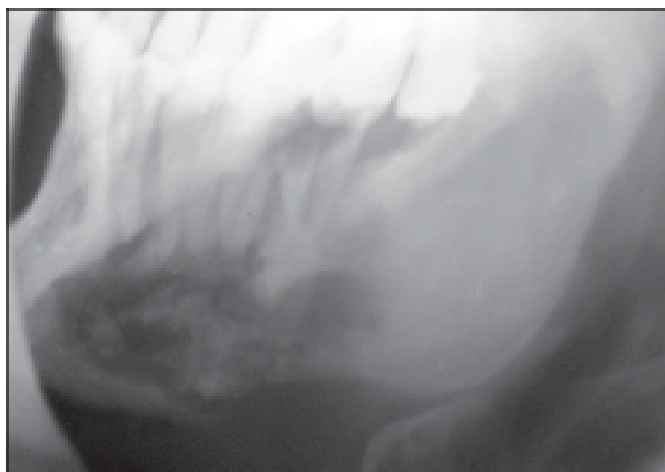


Fig. 19-35: Osteomyelitis with pathological fracture.

Diagnosis

- *Clinical diagnosis*—pain, swelling, fever is present.
- *Radiological diagnosis*—loss of lamina dura, saucer shaped destruction, sequestrum formation and moth eaten appearance will give clue to the diagnosis.
- *Laboratory diagnosis*—the medullary spaces are filled with inflammatory exudate that may or may not progress to the actual formation of pus. The inflammatory cells are chiefly neutrophilic polymorphonuclear leukocytes, but may show occasional lymphocytes and plasma cells.
- *Investigation to be carried out in osteomyelitis*—investigation like gram staining, culture and sensitivity, WBC count and complete hemogram, blood sugar, Mantoux test, radiographs, scintigraphy and computerized tomography.

Differential Diagnosis

- *Paget's disease*—it affects multiple bones and the complete involvement of individual bone.

- *Eosinophilic granuloma*—margins are better than osteomyelitis and have no evidence of bone sclerosis.
- *Difference between carcinoma and osteomyelitis.*

Features	Osteomyelitis	Carcinoma
Age	Usually under 40 years	Post middle age
Progression	Fast	Slower
Extent of involvement	Large involvement and sporadic occurrences	Relatively continuous destruction
Sequestrum	Yes	None
Osteosclerotic formation	Yes	None
Periosteal formation	Large area involved mainly in the parallel direction	Generally none except in some sarcoma
Residual small bony fragment	None	Formation seen within the lesion
Bone scintigram	Negative uptake at the sequestration	Strong positive uptake seen

Management

The goal of definitive therapy is to attenuate and eradicate the proliferating pathogenic microorganisms and to support healing. This is accomplished by removing pathogenic suppurative debris, providing regional stability and disrupting pathophysiology barriers while re-establishing vascular permeability to the infected area.

- *Incision and drainage*—when early diagnosis is made, drainage of the fluctuant areas should be carried out under antibiotic cover. After pus is evacuated, drains are placed. The consistency, color and odor of the pus may provide important clues to the diagnosis and initial treatment.
- *Irrigation and debridement of necrotic areas*—thorough debridement of the affected areas should be carried out. Debride any foreign bodies, necrotic tissue or sequestra. These areas may be irrigated with hydrogen peroxide and saline.
- *Empiric therapy:*
 - *Regimen I*—aqueous penicillin 2 million units, IV, 4 hourly plus oxacillin 1 gm, IV, 4 hourly.
 - *Regimen II*—if the patient is asymptomatic after 48 to 72 hours, then penicillin V 500 mg, 6 hourly and dicloxacillin 250 mg, 4 hourly, for an additional 2 to 4 weeks.
- *Definitive therapy*—culture and sensitivity testing is performed, and the antibiotic therapy is then modified accordingly, particularly if the infection appears to be refractory after the treatment is instituted. If favorable response occurs, no change is indicated.

- *Initial therapy with gram stain result*—if the stain shows predominance of gram -ve cocci in clumps suggestive of staphylococcus, penicillinase resistant penicillin alone is advised.
 - An initial course of oxacillin 1gm IV after every 4 hours.
 - If improvement occurs then orally dicloxacillin 250 mg after every 4 hours should be administered.
- If the stained slide shows predominately gram +ve rods, then an established working diagnosis of anaerobic infection (bacteroides) is made.
 - An aqueous penicillin 2 million units may be given IV, after every 4 hours.
 - An oral antibiotic like penicillin V may be started after 48 to 72 hours.
- *If patient is allergic to penicillin:*
 - *Clindamycin* is recommended. In acute phase 600 mg, after every 6 hours should be given. Once the acute phase is over, orally 450 mg may be given, after every 4 hours.
 - *Cephalosporin* or cefazolin is the third drug of choice in case of allergy to penicillin. Cefazolin 500 mg, IV / IM, after every 8 hours. Once acute symptoms have subsided, cephalexin 500 mg, after every 6 hours is recommended.
 - As a fourth choice *erythromycin* is also useful 2 gm IV after every 6 hours. When acute stage is resolved then 500 mg is given orally, after every 6 hours.
- *Other modalities*—intra-arterial antibiotic therapy and local implantation of antibiotics. Another unique pharmacological treatment is the use of prostaglandin inhibiting salicylates therapy, in an attempt to diminish osteolytic destruction in osteomyelitis model.
- *Extraction*—extraction of carious teeth with periapical infection, should be done. It should be carried out to remove the source of infection from the oral cavity.
- *Supportive therapy:*
 - *Adequate rehydration*—patients is suffering from osteomyelitis required adequate rehydration in the form of fluids.
 - *Rich nutritional diet*—rich nutritional diet should be given.
 - *Vitamin therapy*—multivitamin supplements should be given.
- *Sequestrectomy*—it is the removal of sequestra which are small pieces of necrotic bone that are avascular and harbor microorganisms. Sequestra are not generally seen until two weeks after initiation of the infection. They get resorbed or get spontaneously expelled out from the mucosal or skin surface. If sequestra are not removed, they get chronically infected and also infect the surrounding tissues. This will lead to further sequestration and bone loss. Hence, sequestra need to be removed.
- *Saucerization*—it means excision of the margins of necrotic bone. This will results in better visualization of sequestra and excision of margins of the affected bone. In chronic osteomyelitis, it allows removal of formed and forming sequestra. This usually performed under local anesthesia by raising buccal flap. All the granulation tissue, along with loose bony fragments, is removed from the bony bed using curette and the area is thoroughly irrigated.
- *Closed wound irrigation and suction*—after intraoral sequestrectomy and saucerization, small pediatric nasogastric feeding tubes are used. Drains are placed through separate skin incisions along the decorticated surface and affixed to each other and to the bone with catgut sutures, through the holes drilled in the bone. After tube placement, water tight closure of the wound is achieved and the tubes are flushed with saline. The irrigating solution is flushed through one tube and the other tube is connected to low pressure suction. Various irrigating solutions may be employed, which often contain antibiotics, wetting agents and proteolytic enzymes. Systemic antibiotics should be continued throughout the period of irrigation.
- *Decortication*—decortication of mandible refers to removal of the chronically infected and inferior cortical plates, 1-2 cm beyond the area of involvement. Thus, access is provided to the medullary cavity. Decortication should be performed in sub-acute and chronic stages. It is based on the principle that the involved cortical bone is avascular and harbors microorganisms, while an abscess exists within the medullary cavity where antibiotics can not penetrate. This should be performed where initial conservative regimens have failed.
- *Resection and immediate reconstruction*—resection of the region of osteomyelitis with immediate reconstruction is controversial. Osteotomy is performed in the bleeding bone by intraoral route. Resection is indicated when antibiotics combined with decortication fail to cure the condition in patient where involucrum formation is inadequate, no involucrum in formed after months of antibiotics therapy, the lingual and buccal plates of the mandible are extensively destroyed and considerable destruction of the mandibular ramus occurs without new bone formation. Immediate reconstruction offers the obvious advantage of shortening the period of illness and seeding of rehabilitation.
- *Hyperbaric oxygen therapy*—the scientific foundation for oxygen therapy was laid down in 1960. It was first used in the treatment of gas gangrene. Over the last few decades, hyperbaric oxygen therapy has emerged as a potent alternative to surgical reperfusion and as an adjunctive to host response. HBO therapy has specific applications in oral and maxillofacial surgery; it is used

in the prevention and adjunctive treatment of osteomyelitis, osteoradionecrosis and certain soft tissue infections. The partial pressure of oxygen in osteomyelitis bone is significantly decreased, compared with normal bone. The oxygen tension of normal bone is between 40 to 45 mm of Hg. The oxygen tension in osteomyelitis bone involves a combination of factor, but its oxygen tension in osteomyelitis bone varies from 10-20 mm of Hg. In osteomyelitis, bone has increased consumption of oxygen may be because of greater inflammatory response, cellular exudation and respiration of phagocytes. Secondary to infection, infective organisms also consume more oxygen.

- **Rationale behind oxygen therapy**—adequate tissue oxygen is necessary for fibroblast proliferation. Fibroblast lay down collagen, which provides a framework for capillary formation. The most important aspect of HBO is its positive enhancement of neoangiogenesis in the aerobic portion of the proliferative phase of wound healing. Under the influence of hyperbaric oxygen therapy, endothelial proliferation is seen, which obliterates avascular and ischemic pieces. Free oxygen radical is formed during hyperbaric oxygen therapy that is toxic to many pathogenic anaerobes. HBO is bactericidal for many anaerobic microorganisms. Besides these, the endotoxin liberated by pathophysiological activity of microorganisms is rendered inert by exposure to elevated partial pressure of oxygen. HBO enhances lysosomal degradation potential, by enhancing the development of intracellular halides in leukocytes and oxygen radicals, which are a major component of catabolic enzymes of the macrophage lysosomes. Formation of this enzyme is depressed or decreased in hypoxic environment, such as that found in osteomyelitis foci. Thus, leukocytic bactericidal ability is enhanced. Patient usually experiences relief from pain within 3 to 5 sittings treatment of with rapid closure of draining fistulae. Sequestra get localized and there is rapid progression to osseous reconsolidation and fibrous union.
- **Procedure**—scrubbing the exposed bone with antiseptic solution such as acriminoacridine. Purulent wound should be covered with neomycin dressing. An antibiotic, preferably penicillin, erythromycin or tetracycline, is administered orally. Vitamin E is also given (100 gm) daily to decrease oxygen seizure susceptibility. Patient is placed in multiplace/monoplace chamber and a concentration of 100% O₂ is given. Duration of treatment is of 1.5 to 2 hours for 5 to 6 days in a week, for a total of 60 treatments. Daily wound irrigation and debridement are often required. After 6 months ten additional 2 hour

treatments are advised, followed by ten treatments annually thereafter.

- **Contraindication**—hyperbaric oxygen therapy should not be given in pneumothorax, asthma, COAD, optic neuritis, acute viral infection, URI, congenital spherocytosis, and pregnancy.
- **Complication**—complication of hyperbaric oxygen therapy includes decompression sickness, eustachian tube dysfunction, high pressure nervous syndrome and pneumothorax.

Different Types of Osteomyelitis

Infantile Osteomyelitis

It is a rare type of osteomyelitis seen in infants few weeks after the birth. It usually involves the maxilla.

Route of infection

- **Hematogenous route**—infantile osteomyelitis is usually transferred through the hematogenous route.
- **Trauma**—prenatal trauma of oral mucosa from obstetrician's finger.
- **Infection**—infection from mucous bulb are used to clear the airway immediately after birth.
- **Infected nipple**—infected human or artificial nipple.

Clinical features

- **Sites**—it is more common in maxilla due to hematogenous route. The infection is thought to arise in maxillary antrum or lacrimal sac. It appears to center in the region of first deciduous molars and the adjacent portion of maxilla, although it may involve the inferior aspect of the orbit.
- **Symptoms**—there is fever, anorexia and dehydration. In some cases, convulsions and vomiting may occur.
- **Signs**—redness and edema of eyelids, alveolar bone and palate of the affected side. Intracanthal swelling, palpebral edema, conjunctivitis and proptosis may result. Maxilla on affected side is swollen both buccally and palatally, especially in the molar region.
- **Sinus**—sinus develops and discharges pus intraorally and extraorally.
- **Complication**—complications may occur like TMJ infection and devitalization of adjacent tooth germs may occur.

Radiographic features

- *It will be same as in acute osteomyelitis.*

Diagnosis

- **Clinical diagnosis**—sinus, fever, pain with maxillary involvement in the infant will give clue to the diagnosis.

Management

It is same as that for acute osteomyelitis.

Diffuse Sclerosing Osteomyelitis

In this type, there is reactive proliferation of bone to infectious process occurs. It occurs due to low grade infection. In this, chronic bacterial infection of bone creates smoldering mass of chronically inflamed tissue which initiates sclerosis of surrounding bone.

Clinical features

- *Age and sex*—it can occur at any age but most common in older persons. Most cases are reported in blacks and in female.
- *Sites*—it is common especially in edentulous mandibular jaw.
- *Symptoms*—symptoms are very mild to absent. During the period of growth, the patient complains of pain and tenderness. Pain persists for few weeks to months to even years.
- *Signs*—jaws may be slightly enlarged on the affected side.

Radiographic features

- *Location*—usually large portions of mandible are involved. Surface of new bone is usually smooth.
- *Radiodensity*—there is presence of osteolytic and osteosclerotic zones. As the lesion progresses, there is increase in the size of the involved part.
- *Margins*—margins are ill defined.
- *Granular densification*—stripped or granular densification of bone, caused by subperiosteal deposition of new bone, obscures the intrinsic bone structure or deposition of new bone on the surface of marrow spaces. The deposition is more on the buccal and inferior surface of the jaw.
- *Tooth root*—there is shortening of tooth root.
- *Scintigraphy*—scintigraphy shows uptake of ^{99m}Tc polyphosphate in diseased area, suggesting active bone deposition.

Diagnosis

- *Clinical diagnosis*—as symptoms of this disease are mild, it is not possible to make confirm clinical diagnosis.
- *Radiological diagnosis*—there is granular densification, osteolytic and osteoblastic zone. Scintigraphy will show active bone deposition.
- *Laboratory diagnosis*—there is increased ESR. Biopsy shows bone in 'mosaic' pattern, indicative of repeated periods of resorption followed by repair. The soft tissue between the individual trabeculae is fibrous and show proliferative fibroblasts and occasional small capillaries focal collections of lymphocytes and plasma cells.

Management

- *Infection control*—the best management method for diffuse sclerosing osteomyelitis is resolution of active foci of

infection in the oral cavity. This can be done by extraction of teeth, periodontal management.

- *Management of secondary osteomyelitis*—in some patient, secondary osteomyelitis may develop. It is managed same as that acute and chronic osteomyelitis.

Condensing Osteitis or Focal Sclerosing Osteomyelitis

If the exudate is of low toxicity and long standing then the resulting mild irritation may lead to circumscribed proliferation of periapical bone, appearing as condensing osteitis or focal sclerosing osteomyelitis. There is deposition of new bone along the existing trabeculae, a process known as appositional bone deposition.

Clinical features

- *Age*—it occurs almost in young person, before the age of 20 years.
- *Location*—the tooth commonly affected is mandibular first molar with a large carious lesion. It is associated with non-vital teeth or in teeth undergoing the process of degeneration.
- *Symptoms*—tooth is usually asymptomatic. But in some cases, patient may report pain or tenderness on percussion or palpation.

Radiographic features

- *Appearance*—it appears as localized area of radiopacity surrounding the affected tooth, which may extend below the apex (Fig. 19-36).
- *Margins*—it may be variable in size and extent, with margins from well defined to very diffuse. At the diffused margins, the thickened trabeculae can be seen in continuation of adjacent normal bone (Figs 19-37A and B).
- *Alveolar bone*—the alveolar bone may be sclerose between two adjacent teeth, extending to the crest without any evidence of periodontal disease.

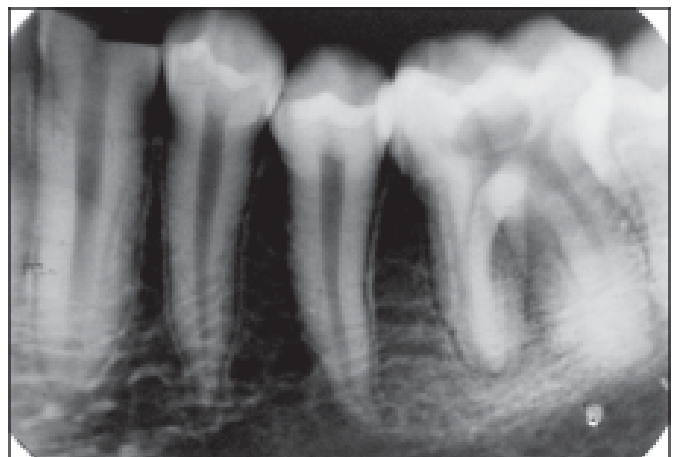
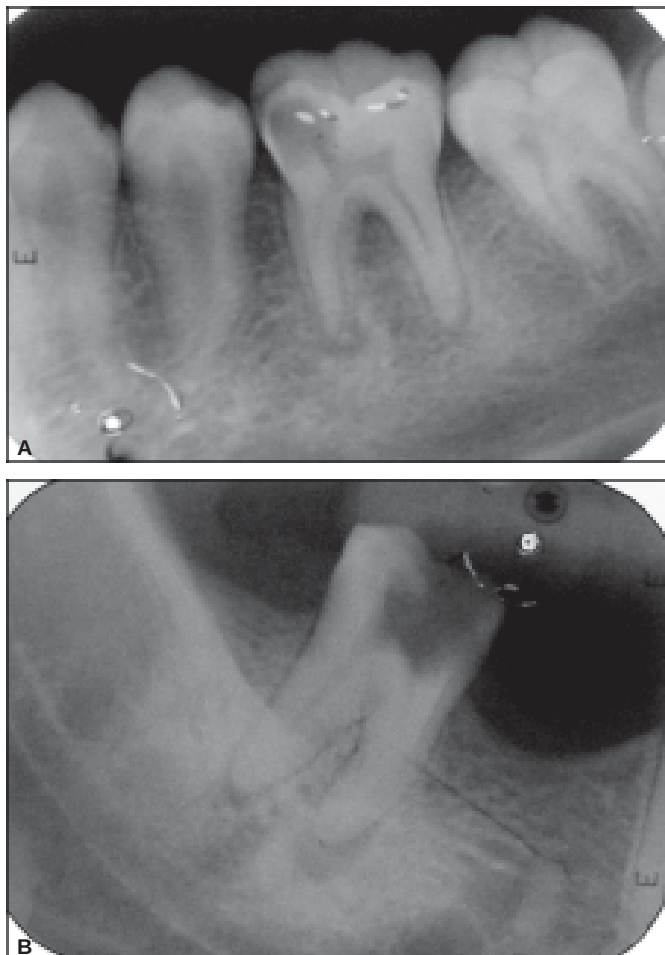


Fig. 19-36: Condensing osteitis seen at the apex of the tooth.



Figs 19-37A and B: Condensing osteitis showing radiopacity at the apex of tooth (Courtesy Dr Ashok).

- *Inferior dental canal*—due to sclerosis, inferior dental canal may be narrowed.
- *Bone scar*—this is the residual area of condensing osteitis after resolution of infection.

Diagnosis

- *Clinical diagnosis*—it is not possible to make clinical diagnosis.
- *Radiological diagnosis*—localized uniform zone of increase radiodensity at the apex of tooth will be diagnosed this condition.
- *Laboratory diagnosis*—it appears as an area of dense bone with trabeculae borders lined by osteoblasts. Chronic inflammatory cells, plasma cells and lymphocytes are seen in the scanty bone marrow.

Differential diagnosis

- *Hypercementosis*—it forms an integral part of the malformed root within intact lamina dura and periodontal ligament space, in contrast to condensing osteitis which is outside the lamina dura and periodontal ligament space.

- *Periapical cemental dysplasia*—it occurs in association with vital tooth and is surrounded by radiolucent halo. Condensing osteitis is not well defined.
- *Osteosclerosis*—it occurs in edentulous area and is associated with vital teeth.
- *Projected radiopacities*—it can be differentiated by 'slob' technique.
- *Enostosis*—it is a small area of greater density that is present in many bones. But in cases of enostosis, more than one are present in the jaws and margins are usually sharp. If the mass is separated from the adjacent tooth by more than couple of millimeters of normal bone, it is enostosis.
- *Odontome*—it has got small radiolucent foci within the dense radiopaque area.
- *Paget's disease*—the adjacent bone also show evidence of altered structure.
- *Torus mandibularis*—the site, bilateral nature and symmetry favors the diagnosis of *torus mandibularis*.
- *Foreign bodies*—there is a related history; the shape and density varies.

Management

- *Endodontic treatment*—endodontic treatment should be carried out with affected tooth. This will resolve the lesion.
- *Extraction*—if the tooth is unrestorable extraction of tooth should be carried out.

Osteomyelitis with Proliferative Periostitis (Garre's Osteomyelitis)

It is also called as '*Osteomyelitis with proliferative periostitis; Periostitis ossificans*'. It was first described by Carl Garre in 1893. But the use of term Garre's osteomyelitis is controversial. The reason for this is that when Garre's discovered it X-ray are not discovered. So the word periostitis does not arise at that time.

It is characterized by formation of hard bony swelling at the periphery of the jaw. It is essentially a periosteal osteosclerosis analogous to the endosteal sclerosis of chronic, focal and diffuse sclerosing osteomyelitis.

For the lesion to develop following conditions should be satisfied.

- The periosteum must possess high potential for osteoblastic activity.
- Mild infection serves as a stimulus.
- Fine balance should be maintained between the resistance of host and number and virulence of organisms.

Clinical features

- *Age and sex*—occurs mainly below 30 years; males are affected more commonly than females.

- **Sites**—most frequently involve the anterior surface of tibia and femur. Mandible is affected more commonly than maxilla. It commonly occurs at the inferior border of mandible, in first molar region.
- **Appearance**—it is presented as hard non-tender swelling with medial and lateral expansion of jaw.
- **Size**—mass varies in size from 1 or 2 cm to the involvement of the entire length on the affected side. Cortex may become 2 to 3 cm thick.
- **Signs**—it may become secondarily infected and cause considerable discomfort.
- **Other findings**—lymphadenopathy, hyperpyrexia and leukocytosis are common findings.

Radiographic features

- **Carious tooth**—intraoral radiograph will reveal a carious tooth opposite the hard bony mass.
- **Cortex**—shadow of thin convex shell of bone over the cortex may be seen. No trabecular pattern between shell of new bone and cortex.
- **Onion skin appearance**—as infection persists, the cortex thickens and becomes laminated (**Fig. 19-38**) with alternating radiopaque and radiolucent layers (*onion skin appearances*).
- **Cancellous bone**—adjacent cancellous bone may remain normal, become sclerotic or show some areas of osteolytic changes within the scleroses spongiosa.
- **Small sequestra**—within the new bone osteolytic radiolucencies or small sequestra are present.
- **Computed tomography**—it is superior to conventional radiography.

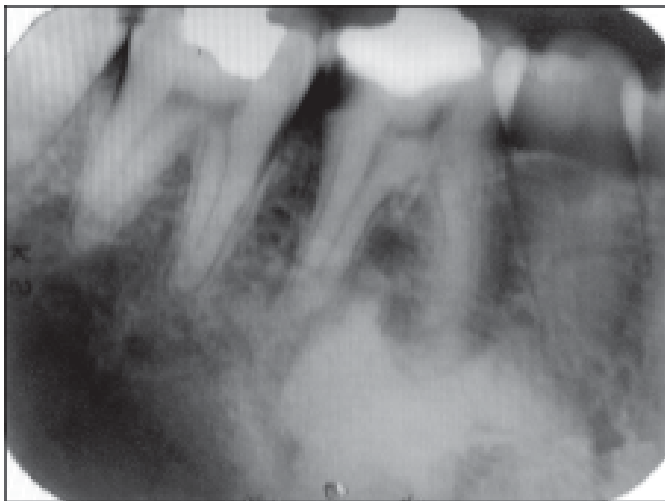


Fig. 19-38: Proliferative osteomyelitis showing radiopacity at the apex which is having laminar structure.

Diagnosis

- **Clinical diagnosis**—it is not possible to make clinical diagnosis.

- **Radiological diagnosis**—'onion peel appearance' is seen as lamination parallel to cortex.
- **Laboratory diagnosis**—biopsy shows reactive new bone and osteoid tissue, with osteoblasts bordering many of the trabeculae. These trabeculae often are oriented perpendicular to the cortex, with the trabeculae arranged parallel to each other or show a retiform pattern.

Differential diagnosis

- **Ewing's sarcoma**—Bony enlargement caused by sarcoma develops more rapidly. Sun ray appearance is classical. Facial neuralgia and lip paresthesia are frequent complications of this entity.
- **Caffey's disease** (infantile cortical hyperostosis)—It is found at the angle or ramus of the mandible and presents before 2 years as compared to Garre's osteomyelitis which is found in posterior tooth bearing areas with later onset. Caffey's disease is developed in more than one bone, as compared to Garre's osteomyelitis, which is developed in one bone.
- **Fibrous dysplasia**—it is not associated with dental infection and is likely to be found in maxilla. In Garre's osteomyelitis, there is thickened cortex, as compared to fibrous dysplasia in which there is a thin cortex. Density of fibrous dysplasia is much more uniform than in Garre's osteomyelitis, which may be mottled with alternating areas of sclerosis and osteolysis.
- **Osteosarcoma**—sunray appearance is seen.
- **Ossifying sub-periosteal hematoma**—it will not be so uniformly radiopaque, having either more mottled appearances or even trabeculated pattern. History of trauma is present.
- **Peripheral osteoma, tori, exostosis**—they appear as dense uniform radiopaque masses on the jaw or protruding from the cortex.
- **Infantile cortical hyperostosis**—generalized expansion of cortices of several bones.

Management

- **Endodontic therapy**—this should be carried out in the tooth.
- **Extraction**—if the tooth is unrestorable then extraction should be carried out.

Actinomycotic Osteomyelitis

Actinomycosis is a chronic infection manifesting, both, granulomatous and suppurative features, that usually involve the soft tissues and occasionally the bone of cervicofacial, abdominal and thoracic region. About 1/3rd of the cases are cervicofacial.

Patient presents with soft tissue masses of skin, which have purplish, dark red, oily areas with occasional zone of fluctuance. Actinomycosis produce *lumpy jaw*—actinomycosis infection produces dense bone and scarring

of soft tissues and has been described as 'lumpy jaw'. Actinomycosis will result in osteomyelitis of jaw bone. It is common in cattle and rare in human being. Soft tissues provide a harbor for organisms and reduce the blood supply to the affected region.

Tuberculous Osteomyelitis (Fig. 19-39)

Bone and joint tuberculosis is always hematogenous in origin. Primary focus is related to lung when disease is acquired by inhalation of human strain or to gastrointestinal tract if it is acquired by ingestion of bovine tubercle. The disease starts within the synovial membrane or in intra-articular bone. The disease may develop in synovial joints especially the knee and hip joint. Tuberculous osteomyelitis of maxilla or mandible or TMJ are rare entities.

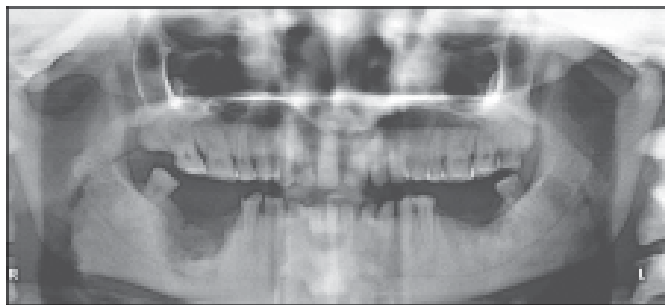


Fig. 19-39: Moth eaten type of appearance seen in mandible due to osteomyelitis with history of tuberculosis in this patient.

Radiation Osteomyelitis

It is an infection of the irradiated bone. It occurs after exposure to radiation (40 to 80 Gy), bone undergoes marked decreased in vascularity. Such bone has poor defensive process and is susceptible to traumatic injury. It draws infection from extraction wound and infected pulp, severe periodontitis and denture stomatitis. It will cause death of bone cells and result in progressive obliterative arteritis. This results in aseptic necrosis of the portion of bone directly in beam of radiation, with compromised vascularity in the adjacent bone.

Pathogenic organisms are introduced into this irradiated bone through odontogenic infections, compound fractures of the jaws and mucosal lacerations. The organisms most commonly found are *Staphylococcus aureus*, *Staphylococcus epidermidis*. Higher incidences in jaw are necrotic due to higher degree of infections and frequent trauma to which these bones.

Osteomyelitis can occur in irradiated jaw. Intense pain and facial fistula develop from the subperiosteal tissues. Spread is diffuse and throughout with signs of inflammation and swelling.

Chronic Tendoperiostitis

It is reactive hyperplasia of bone that is exacerbated by overuse of the masticatory muscle. It is caused by parafunctional habits like bruxism, clenching, nail biting. Masseter inhibitory reflexes were abnormal in this case.

Clinical Features

- *Age and sex distribution*—it is commonly seen in 4th decade of life without sex predilection.
- *Site*—it is more common in masseter and digastric muscle.
- *Symptoms*—recurrent pain, swelling of cheek, trismus, triad of symptoms.
- *Signs*—there is as such no suppuration is present.

Radiological Features

- *Location*—it is seen in anterior region of mandibular angle and posterior region of mandibular body.
- *Sclerosis*—sclerosis is limited to the one quadrant only. There is radiolucent zone inside the radiopacity.
- *Erosion*—erosion of inferior border of mandible can be present.

Diagnosis

- *Clinical diagnosis*—triad of symptoms of cheek swelling, trismus and recurrent pain will give clue to the diagnosis.
- *Radiological diagnosis*—sclerosis at angle of mandible with radiolucent zone in it is present.
- *Laboratory diagnosis*—biopsy show reactive bone, dense bone with few foci of inflammation.

Management

- *Muscular relaxation method*—this is carried out by rotational exercise, myofeedback, soft diet should be given.
- *Muscle relaxant drugs*—muscle relaxant drugs like diazepam, meprobamate should be given.
- *Occlusal splint*—occlusal splint should be given to the patient to control parafunctional habits.

SAPHO Syndrome

SPAHO syndrome includes Synovitis, Acne, Pustulosis, Hyperostosis, and Osteitis. It occurs due to unknown cause. Genetic predisposition is present in this case.

Clinical Features

- *Age and sex distribution*—patient affected in the age group of 50 to 60 years.

- *Site*—most commonly involve site are anterior chest wall, clavicle, and ribs.
- *Skin lesion*—palmoplantar pustulosis, pustular psoriasis, and acne is present.

Radiological Features

- *Appearance*—osteolytic areas are scattered in the randomly in the sclerotic bone.
- *Periosteal bone reaction*—it is present in many cases. There is no perforation of bone.
- *External bone resorption*—external bone resorption of mandible is common in this condition.
- *Scintigraphy*—it will show involvement of multiple site of bone deposition.

Diagnosis

- *Clinical diagnosis*—it is easy to make clinical diagnosis by 5 typical features of this syndrome.
- *Radiological diagnosis*—periosteal bone reaction with multiple sites of bone deposition.
- *Laboratory diagnosis*—biopsy shows bone remodelling, there is also interconnecting trabeculae of vital reactive mixed with fibrous connective tissue.

Management

- *Steroidal and non-steroidal anti-inflammatory drugs*—these are most effective agents to relieve the symptoms.
- *Decortication*—this is effective in some cases of SPAHO syndrome.

Facial Space Infections

Facial spaces are potential spaces situated between the planes of fascia, which form natural pathways along which the infection may spread.

- *Primary*—these are directly related to teeth:
 - *Maxillary spaces*—it includes canine space, buccal space, infratemporal space and parotid space.
 - *Mandibular spaces*—it includes space for body of mandible, submental space, sublingual space, submandibular space, buccal space and pterygomandibular space.
- *Secondary*—these are not directly related to teeth involved maxillary spaces. It includes:
 - Masseteric
 - Pterygomandibular
 - Superficial and deep temporal
 - Lateral pharyngeal
 - Retropharyngeal
 - Prevertebral.

Based on Clinical Significance

- *Face*—buccal, canine, masticatory, parotid.
- *Suprahyoid*—sublingual, submandibular, pharyngo-maxillary, peritonsillar.
- *Infrahyoid*—anterovisceral (pretracheal)
- *Space of total neck*—retropharyngeal, space of carotid sheath.

Different types of space infection is discussed below:

Canine Space

Anatomy

- *Location*—one of the potential spaces that is frequently the seat of infection is the canine fossa. This space actually lies between the anterior surface of the maxilla and the overlying elevator muscle of upper lip.
- *Content*—the fossa contains considerable amount of connective tissues and fat, which allow the accumulation of tissue fluid and pus.
- *Boundaries*—it is bounded by:
 - *Superiorly*—by levator labii superioris alequae nasi.
 - *Anteriorly*—by orbicularis oris.
 - *Posteriorly*—by buccinator.

Clinical Features

- *Location of swelling*—swelling just lateral to the nose, obliterating the nasolabial fold. In some cases canine space infection may extend to orbit (**Fig. 19-40**).
- *Intraoral location*—intraorally, swelling is present in the labial sulcus. Rarely, palatal swelling is encountered.



Fig. 19-40: Canine space infection extending to the orbit.

Buccal Space

Anatomy

- **Location**—this space is situated between the buccinator and masseter muscle. It lies superficial to the buccinator muscle and buccopharyngeal space.
- **Contents of space**—it contains buccal pad of fat, Stensen's duct, anterior facial artery and transverse facial artery and vein.
- **Boundaries**—it is bounded by:
 - *Medially*—by buccinator muscle and buccopharyngeal fascia.
 - *Laterally*—by skin of cheek.
 - *Anteriorly*—by zygomatic arch and depressor muscle of corners of mouth.
 - *Superiorly*—by zygomatic arch.
 - *Inferiorly*—by lower border of mandible.
 - *Posteriorly*—by masseter muscle and pterygomandibular raphe.

Clinical Features

- **Location of swelling**—swelling extent from lower border of mandible to level of zygomatic arch (**Fig. 19-41**). Swelling arises due to infection from mandibular or maxillary molar or premolar, if apex is above the attachment.
- **Symptoms**—facial swelling with little trismus.
- **Sign**—swelling is dome shaped and in some cases periorbital edema develops.



Fig. 19-41: Buccal space infection showing dome shape swelling.

Parotid Space

Anatomy

- **Content**—it is a compartment formed by splitting of investing layer of deep cervical fascia. It contains parotid gland as well as extra-glandular and intra-glandular

parotid lymph nodes. It also contains external carotid, internal maxillary and superficial temporal artery.

- **Attachment of gland**—the gland is itself attached strongly to the facial covering and there is very little intervening connective tissue. This makes extension of infection into parotid space very difficult.

Origin of Infection

- **Blood born infection**—most of the infections are blood borne or occur as retrograde infection through the parotid duct.
- **Retrograde extension**—it reaches from the lateral pharyngeal space or by retrograde extension along the parotid gland.
- **May break into lateral pharyngeal space**—primary infections of parotid space break into lateral pharyngeal space readily because the fascia is thin over the deep portion of the parotid space.

Clinical Features

- **Location of swelling**—the swelling extends from the level of the zygomatic arch to the lower border of the mandible. Posteriorly, it extends into the retromandibular region and anteriorly, it ends at the end of the anterior border of ramus.
- **Signs**—the swelling tends to evert the lobule of ear.
- **Symptoms**—the patient complains of pain, which is referred to the ear and accentuated on eating.
- **Diagnosis**—diagnosis is made by following features:
 - Eversion of lobule of ear (**Fig. 19-42**).
 - No evidence of trismus.
 - Possible escape of pus from parotid duct, when the gland is milked.
 - All signs of abscess formation.



Fig. 19-42: Parotid space infection showing eversion of ear lobule.

Infratemporal Space

Anatomy

- **Location**—it is an irregularly shaped space which lies behind the posterior surface of the maxilla.
- **Contents of space**—it contains pterygoid plexus, internal maxillary artery, the mandibular, mylohyoid, lingual, buccinator and *chorda tympani* nerves and external pterygoid muscle.
- **Boundaries**—it is bounded by:
 - **Laterally**—by tendon of the temporal muscle, coronoid process and ramus of mandible.
 - **Medially**—by lateral plate of pterygoid process, inferior belly of the lateral pterygoid muscle and lateral wall of pharynx.
 - **Posteriorly**—by lateral pterygoid muscle, condyle, temporal muscle and the fossa is limited by parotid gland, which overlaps into it.
 - **Anteriorly**—it is limited by maxillary tuberosity.
 - **Superiorly**—the roof of infra-temporal fossa is formed by greater wing of sphenoid.
 - **Inferiorly**—it communicates with pterygomandibular space.

Spread

- **Spread to cavernous sinus**—infra-temporal infection can extend via the plexus of veins, through the inferior orbital fissure into the terminal part of inferior ophthalmic vein and then, through the superior orbital fissure into cavernous sinus.
- **Spread to pterygomandibular space**—the infection from this space can extend into pterygomandibular space.

Clinical Features

- **Location**—extraoral swelling over the region of sigmoid notch and TMJ joint area and intraorally swelling is usually in the tuberosity area.
- **Symptoms**—the patient may exhibit trismus and sometimes swelling of eyelids, if there is involvement of post-zygomatic fossa.
- **Signs**—entire cheek may be swollen, if the buccal space is also involved.
- **Pharynx involvement**—the involvement of pharynx may cause dysphagia and severe pain or a feeling of pressure in the general area of infection.

Space for the Body of Mandible

Anatomy

- **Location**—it is formed as the external cervical fascia splits medially and laterally, at the inferior border of the mandible and becomes continuous superiorly with alveolar mucoperiosteum.

- **Content**—it contains the mandible anterior to the ramus, various mandibular attachments blood vessels, nerves and periodontal structures. The firm attachment of the mandibular periosteum at the inferior border serves to prevent the extension of infection inferiorly into the neck.

Origin

- **Fracture or direct extension**—infection may arise from fracture or by direct extension from the floor of mouth, lateral pharyngeal spaces and masticator space.
- **Dental, periodontal or blood born**—infection may be dental, periodontal or vascular in origin.

Clinical Features

- **Location of swelling due to infection from incisor, canine and cuspid teeth**—swelling can be present in different location, according to origin of infection. Infection from incisors, cuspid or bicuspid teeth also can cause infection of space of the body of mandible.
 - **Outer cortical plate involvement**—if outer cortical plate is involved then there is induration or fluctuation of the labial sulcus.
 - **Inner cortical plate involvement**—when the inner cortical plate is involved, the infection is restricted to the floor of mouth (**Fig. 19-43**).



Fig. 19-43: Swelling seen floor of mouth due to space for body of mandible infection

- **Location of swelling due to infection from molar teeth:**
 - **Perforation of infection above the external oblique ridge**—if the infection perforates the bone above the external oblique, swelling in the oral vestibules occurs.
 - **Perforation below the mylohyoid line**—if the perforation is below the mylohyoid line the infection may point in the skin.

Submental Space

Anatomy

- *Location*—this space lies in the midline between *symphysis menti* and the hyoid bone.
- *Content of space*—it contains submental lymph nodes, which drain the median part of lower lip, tip of tongue and floor of mouth.
- *Boundaries*—it is bounded by:
 - *Floor*—its floor is formed by the mylohyoid muscle.
 - *Roof*—its roof is formed by suprahyoid portion of investing layer of deep cervical fascia.
 - *Lateral*—laterally by anterior belly of digastric muscle.

Clinical Features

- *Location of swelling*—chin becomes grossly swollen, quiet firm and erythematous.
- *Symptoms*—in some cases it may cause dyspnea and dysphagia.
- *Signs*—slight extraoral swelling just below the chin is evident (Fig. 19-44).



Fig. 19-44: Submental space infection showing swelling below the chin

Submandibular Space

Anatomy

- *Location*—it lies lateral to submental space.
- *Contents of this space*—it contains superficial part of submandibular salivary gland and its lymph nodes, the facial artery deep to the gland, proximal portion of the Wharton's duct, the lingual and hypoglossal nerve as they course deep to the gland and facial vein superficial to the gland.
- *Boundaries*—it is bounded by:

- *Laterally* by submandibular skin, superficial fascia, platysma muscle, superficial layer of deep cervical fascia and lower border of mandible.
- *Medially*—by mylohyoid, hyoglossus and styloglossus muscle.
- *Inferiorly*—by anterior and posterior bellies of digastric muscle.
- *Posteriorly*—the space extends to the hyoid bone.

Origin

Odontogenic infection of this space is caused by 2nd and 3rd mandibular molars.

Clinical Features

- *Location of swelling*—it produces a swelling near the angle of the jaw (Fig. 19-45).
- *Signs*—swelling is brawny edematous in appearance. After some days, swelling becomes soft and fluctuant.
- *Gland and lymph node involvement*—as this space lies in close proximity with submandibular gland and lymph node there is always chance of involvement of gland and lymph nodes. This will result in sialadenitis and lymphadenitis.



Fig. 19-45: Submandibular space infection showing swelling at angle of mandible

Sublingual Space

Anatomy

- *Location*—it lies above the mylohyoid muscle.
- *Contents of space*—it contains sublingual gland, submandibular duct, deep portion of the submandibular gland, lingual and hypoglossal nerves and terminal branches of lingual artery.
- *Boundaries*—it is bounded by:
 - *Superiorly*—its roof is formed by mucous membrane of the floor of mouth.
 - *Anteriorly and laterally* by inner surface of the body of mandible above the mylohyoid line.

- *Medially*—by geniohyoid, genioglossus muscle and median raphe of tongue.
- *Posteriorly*—by hyoid bone.
- *Inferiorly*—the floor is formed by mylohyoid muscle.

Origin

The infection may arise directly from perforation of the lingual cortical plate, above the mylohyoid attachment or by extension from other spaces, primarily submandibular space.

Clinical Features

- *Location of swelling*—brawny erythematous tender swelling of the floor of mouth is present. Swelling is close to mandible and spreads towards midline or beyond (Fig. 19-46).
- *Symptoms*—elevation of the tongue may be noted in late cases. Dysphasia and dyspnea may be the related complains.



Fig. 19-46: Sublingual space infection showing swelling in floor of mouth

Submasseteric Space

Anatomy

- *Location*—it includes sub-periosteal region of mandible and facial sling, containing ramus of mandible and muscles of mastication. This space is formed by splitting of the investing layer of deep cervical fascia. The splitting occurs as the fascia is attached to the lower border of the mandible.
- *Content of space*—it contains muscles of mastication, internal maxillary artery and mandibular nerve.
- *Boundaries*—it is bounded by:
 - *Anteriorly* by body of mandible.
 - *Posteriorly* by parotid space.
 - *Medially*—lateral pharyngeal space.

- *Superiorly*—it is continuous with superficial and deep temporal pouches.

Clinical Features

- *Location of swelling*—the swelling may be either external or internal, or both.
- *External swelling*—the external swelling consists of brawny induration over the ramus and angle of mandible (Fig. 19-47).
- *Internal swelling*—internal swelling may predominate in some cases. Such swelling involves the sublingual region and the pharyngeal wall. The pharyngeal swelling pushes palatine tonsils towards midline.
- *Symptoms*—pain is excruciating and often radiates to the ear. Dysphagia may be present, especially when the swelling is internal.
- *Trismus*—clinically masticatory space infection is likely to be marked by severe trismus because of irritation of masseter and medial pterygoid. It can be so severe that the mouth opening is restricted only to half a centimeter.
- Lateral pharyngeal wall, behind the palatine tonsil, is not swollen, which will help to differentiate between it and lateral pharyngeal space infection.



Fig. 19-47: Submasseteric infection showing swelling over the ramus of mandible.

Temporal Space

Anatomy

- *Location*—these are the facial spaces in relation to temporalis muscle. They are two in number, i.e. superficial and deep.

- *Superficial temporal space* lies between the temporal fascia and temporalis muscle.
- *The deep temporal pouch* lies deep to the temporalis muscle, between the latter and the skull.
- *Content*—it contains pterygoid plexus, internal maxillary artery, mandibular nerve and external pterygoid muscle.
- *Boundaries*—it is bounded by:
 - *Anteriorly*—it is bounded by maxillary tuberosity.
 - *Posteriorly*—it is bounded by lateral pterygoid muscle, condyle, and temporal muscle.
 - *Laterally*—lateral pterygoid plate, and inferior belly of lateral pterygoid muscle.

Clinical Features

- *Location of swelling*—in case of infection with superficial temporal space, swelling is limited below by the zygomatic arch and laterally, it is limited by the outline of superficial and temporal line (**Fig. 19-48**). A deep temporal abscess produces less swelling than one involving the superficial temporal space. Since it lies deep to temporalis muscle, fluctuance is difficult to elicit.
- *Dumble shaped appearance*—when it is associated with buccal space infection, the swelling has a characteristic *dumble shaped appearance* caused by the lack of swelling over the zygomatic arch.
- *Symptoms*—due to limited distensibility of temporal fascia, the pain is severe.
- *Trismus*—it is a common finding.



Fig. 19-48: Infratemporal space infection showing swelling zygomatic arch area.

Pterygomandibular Space

Anatomy

- *Location*—it is a well defined space between the mandibular ramus and pterygoid muscle.
- *Contents of space*—it contains variable amount of fat, inferior alveolar nerve and maxillary artery.
- *Boundaries*—it is bounded by:
 - *Lateral wall*—its lateral wall is formed by inner surface of ramus of mandible.
 - *Medial wall*—its medial wall is formed by medial pterygoid muscle.
 - *Roof*—its roof is formed by inferior head of lateral pterygoid muscle.
 - *Frontal section*—in frontal section the pterygomandibular space is a triangular space narrowing downward, where medial pterygoid converges with the mandible.
 - *Posterior*—posteriorly, with retromandibular space containing parotid gland.
 - *Anterior*—anteriorly, the pterygomandibular space is accessible between the deep tendon of temporal muscle, which is attached to the temporal crest of the mandibular ramus and anterior border of pterygoid muscle.

Clinical Features

- *Location of swelling*—there is no external evidence of swelling. Intraoral examination reveals an anterior bulging of half of the soft palate.
- *Signs*—deviation of tongue to the affected side.
- *Symptoms*—severe trismus and difficulty in swallowing.

Lateral Pharyngeal Space

It is also called as '*parapharyngeal space*', '*pharyngomaxillary*' or '*pterygopharyngeal space*'.

Anatomy

- *Location*—it is a lateral neck space whose shape is that of an inverted cone, with the base at the skull and its apex at the hyoid bone. Its medial wall is continuous with the carotid sheath and lies deep to the pharyngeal muscle. It is situated between the lateral wall of pharynx and fascia covering the pterygoid muscle.
- *Contents of space*—it contains fat, muscles and lymph nodes, connective tissue, cranial nerve IX and XII, carotid artery, internal jugular vein and vagus nerve.
- *Boundaries*—it is bounded by:
 - *Laterally*—by carotid sheath, internal pterygoid muscle and mandibular ramus.

- *Medially*—by lateral wall of pharynx and stylohyoid muscle.
- *Anteriorly*—by buccopharyngeal aponeuroses, parotid gland and external pterygoid muscles. It is limited by pterygomandibular space.
- *Posteriorly*—by prevertebral fascia. It communicates with retropharyngeal raphe.

Origin

The source of infection is 3rd molars and sometimes, 2nd molars, particularly by way of infection in the submandibular space or by direct extension the tooth.

Clinical Features

- *Location of swelling*—if the infection is confined to the anterior compartment, external swelling occurs anterior to the sternocleidomastoid muscle. The swelling is first seen at the angle of mandible and in the submandibular region.
- *Symptoms*—there is severe pain on the affected side of throat. Pain is sometimes referred to the ears. Infection of space occurs with abscess formation, which may impinge on pharynx and result in pain while swallowing and making it impossible. Patient also complaint of trismus irritation of medial pterygoid muscle.
- *Signs*—internally, the anterior part of lateral pharyngeal wall is swollen and it pushes the palatine tonsils together, with the soft palate towards the mid line.
- *Posterior compartment involvement*—with infection of posterior compartment, the clinical picture is marked by septicemia.
- *Complication*—respiratory paralysis resulting from acute edema of larynx. There is also thrombosis of internal jugular vein and erosion of internal carotid artery. The lateral pharyngeal space communicates with the mediastinum by prevertebral fascia, so the infection may reach to this area as direct extension.

Retropharyngeal Space

Anatomy

- *Location*—it is situated between the posterior walls of pharynx and prevertebral fascia, extending from the base of the skull to mediastinum.
- *Boundaries*—it is bounded by
 - *Laterally*—it is bound by lateral pharyngeal space and on each side by carotid sheath.
 - *Anteriorly*—by the wall of pharynx.
 - *Posteriorly*—by prevertebral fascia.
- *Danger space*—infections in this region are dangerous because of ease of spread to mediastinum and for this reason, this space is termed as danger space.

Origin

Infection result due to medial extension of infection in the lateral pharyngeal space.

Clinical Features

- *Age*—these infections are classically seen in children less than 5 years.
- *Cause*—these are generally due to otitis media and pharyngitis.
- *Symptoms*—patient will have pain, dysphagia and nuchal rigidity.
- *Signs*—there is bulging of posterior pharyngeal wall. Prominence is noted on one side due to adherence of median raphe or prevertebral fascia.
- *Radiographic finding*—it will show widened retropharyngeal space. Computed tomography can be used to diagnose this condition (**Fig. 19-49**).
- *Complication*—infection may be spread to mediastinum, causing mediastinitis.

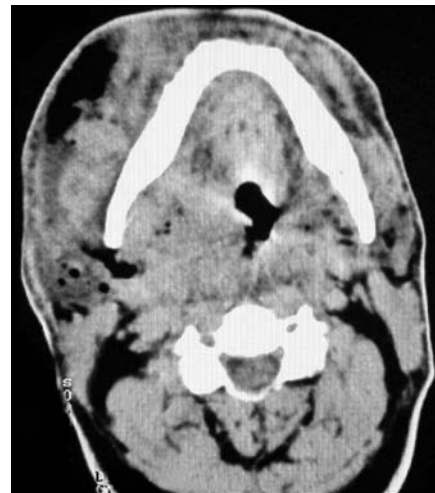


Fig. 19-49: Computed tomography showing air spaces seen in fascial space infection.

Management of Facial Space Infection

- *Medicinal treatment*—it consist of antibiotic and analgesic therapy. Most commonly given antibiotics are penicillins, amoxicillin and ornidazole. In severe cases cephalosporins group antibiotics should be given. Usually combination of antibiotics should be given.
- *Supportive therapy*—adequate hydration, rich nutritional supplements and treatment of pre-existing diseases.
- *Extraction*—extraction of offending tooth should be carried out.
- *Incision and drainage*—surgical evacuation of pus is necessary for two reasons, i.e. to prevent further burrowing of the purulent mass, in an attempt towards spontaneous evacuation and to avoid dreaded

complications like airway embarrassment or erosion of major vessels. Technique for incision and drainage is described below.

- *Preparation of skin*—the skin is prepared in aseptic manner and prepared area is draped with sterile towels.
- *Local anesthesia*—if local anesthetic is used, a ring block of peripheral skin wheel is made for skin anesthesia.
- *Site of incision*—it should be made in the most dependent part of the abscess than in the center where the abscess points through the skin by necrosis of overlying tissue. This provides dependent drainage, avoids puckering of skin and excessive contracture of scar. Hence, it is advised to place incision on healthy skin or mucosa so that tissue heals with flat linear scar, which is more acceptable. Secondly, the incision should be placed in cosmetically and functionally acceptable place.
- *Blunt dissection* is done after an initial sharp incision through skin or mucosa. Blunt dissection by sinus forcep is performed by gentle poking and opening beaks of the instrument, until the abscess cavity is reached. Within the tissue, the beaks of the sinus forceps should never be closed together as it may crush any vital structure. The direction in which the beaks of sinus forcep are spread apart is important. This should be done parallel to the important vital structures so that if they are encountered, they are stretched rather than torn.
- *Dissection*—dissection should be extended to the alveolar process overlying the roots of the involved teeth, i.e. the source of infection.

Fatal Complications of Oral Infection

Bacterial Meningitis

It is the most common neurologic complication resulting from oral and maxillofacial infections. In this condition, bacteria infect arachnoid pia mater and CSF. The infection quickly spreads from its point of origin, via CSF, to the entire subarachnoid space.

Clinical Features

- *Symptoms*—headache, chills, fever and nausea. Pain in back and stiffness of neck.
- *Signs*—Kernig's sign (restriction of knee movement when hips are flexed due to spasm of hamstring) and Brudzinski's sign like neck sign (on flexing neck there is flexion of the hips and knee), leg sign (on flexing one leg,

the other leg also flexes) and cheek sign (pressure against cheek below zygoma will cause reflex flexion at elbow with upward jerking of the arm) are positive.

- *Diagnosis*—diagnosis is made by lumbar puncture and CSF examination.
- *Microorganisms responsible*—*Staphylococcus*, *Streptococcus*, pneumococci, *Proteus*, *Klebsiella* and *H influenzae* are common pathogens causing meningitis secondary to head and neck infection.

Diagnosis

- *Clinical diagnosis*—positive Kernig's and Brudzinski's sign is positive.
- *Laboratory diagnosis*—diagnosis is made by lumbar puncture and CSF examination.

Management

- It is a medical emergency and prompt intravenous antibiotics should be started, after antibiotic sensitivity testing.

Brain Abscess

It can develop from bacteremia associated with odontogenic infections.

Pathogenesis

- Once the bacteria reach the brain tissues →it results in local cerebritis →inflammatory exudate collects along with degenerative leukocytes →septic thrombosis of blood vessels occurs →cerebral edema develops around the infected area →as the abscess grows, its center becomes pus filled with a capsule of granulation tissue at its periphery →as the abscess increases in size, it may rupture into the ventricular system and the result is usually fatal →if it extends to subarachnoid space, meningitis may develop.

Clinical Features

- *Symptoms*—headache is due to rise in intracranial pressure. Nausea, convulsions and vomiting may occur. Dysphagia and visual defects are seen, if it involves the temporal lobe.
- *Signs*—papilloedema and convulsions are common. Hemiparesis, if abscess is on motor cortex. Confusion and stupor, if abscess is in frontal lobe.
- *Diagnosis*—diagnosis is made by radionuclide scanning and CT scanning.

Diagnosis

- *Clinical diagnosis*—papilloedema, convulsion dysphagia can give clue to diagnosis.

- *Laboratory diagnosis*—diagnosis is made by radionuclide scanning and CT scanning.

Management

- *Drainage*—drainage of the abscess via catheter through a bur hole is the treatment of choice. Once the size of the abscess is decreased the complete capsule should be excised.
- *Craniotomy*—some surgeons prefer to do craniotomy initially and then excise the entire abscess.

Cavernous Sinus Thrombosis

It is one of the most dreaded and life threatening complication due to intracranial spread of infection from odontogenic source.

Anatomy

- *Location*—they are paired sinuses situated on either side of sella turcica on lateral slopes of sphenoid bone. Each sinus is an irregularly shaped space within the dura, between the body of the sphenoid bone and medial border of middle cranial fossa.
- *Extent*—it extends from superior orbital fissure to the apex of petrous portion of temporal bone. It has anastomosis anteriorly and inferiorly with pterygoid plexus through foramen of vasallus, foramen ovale and foramen lacerum.
- *Nerve passing through sinus*—the oculomotor and trochlear nerves, the ophthalmic and maxillary divisions of trigeminal nerve passes through the sinus on the lateral wall. The abducens nerve and internal carotid artery, with its surrounding sympathetic plexus, is suspended in the sinus between the lateral wall and sphenoid bone. The blood in the sinus flows around these structures, but are separated from each other by fibrous sheath.

Etiopathogenesis

- *Infection of maxillary premolar and molar teeth*—infection from tooth may perforate buccal cortical plate. Then infection reaches to the maxillary sinus, pterygopalatine space, reaching the orbit via inferior orbital fissure. After this, infection can spread to cavernous sinus at cranial vault and results in cavernous sinus thrombosis.
- *Infection from maxillary anterior teeth*—in this, infection spread to sinus through canine space.
- *Septic emboli*—bacteria can reach cavernous sinus in septic emboli through venous and arterial systems. Septic thrombophlebitis of emissary veins can directly lead to this phenomenon.
- *Infratemporal space infection*—it can also result from direct extension through skull, by an abscess located in deep spaces such as in infratemporal space.

- *Direct antral infection*—the direct antral infection may also give rise to this disease.

Bacteriology

Many organisms have been found in culture of CSF including pseudomonas, corynebacterium, *Staphylococcus albus*, *Streptococcus*, *Diplococcus pneumonia* and *proteus*. Most common organism is *Staphylococcus aureus*.

Clinical Features

- *Symptoms*—there is high spiking fever.
- *Signs*—signs of meningeal irritation including severe headache, stiffness of neck, ocular palsy and facial weakness.
- *Eye*—proptosis or protrusion of eye is seen as a result of decreased venous drainage, chemosis and edema of eyelid, which is secondary to venous stasis.
- *Cranial nerve involvement*—limitation of extraocular movements because of involvement of 3rd, 4th and 6th cranial nerves.
- *Cranial nerve palsy*—cranial nerve palsy of 3rd, 4th and 6th nerve is usually evident due to irritation caused by pressure of venous congestion.
- *Progress*—rapid progression of signs and symptoms from one eye to other eye, due to spread of infection through inter-cavernous sinus.

Diagnosis

- *Clinical diagnosis*—cranial nerve palsy, eye involvement, limitation of extraocular movement will give clue to the diagnosis.
- *Laboratory diagnosis*—lumbar puncture should be made. CSF shows neutrophils, decreased glucose concentration and elevated protein concentration.

Management

- *Antibiotics*—massive doses of antibiotics, with proper surgical intervention at the primary site of infection are essential. The initial drug of choice is IV chloramphenicol 1gm, after every 6 hours. An attempt should be made to identify the causative organism by culture from the source of infection or from blood culture, so that precise antibiotic sensitivity is established.
- *Anticoagulant therapy*—use of anticoagulant therapy to prevent further thrombosis and dissemination of septic emboli. But some investigator think that emboli limit the infection and use of anticoagulant may promote hemorrhagic lesion in orbit and brain.

Odontogenic Infection of Orbit

It can result in significant morbidity and mortality.

Etiology

- *Infection from premolar and molar*—odontogenic infection can spread to orbit through several routes. Infection of premolars and molars can perforate the maxillary buccal plate and can spread to pterygopalatine and infra-temporal fossa and reach the orbit, via inferior orbital fissure.
- *Maxillary anterior teeth*—the maxillary anterior teeth can produce orbital cellulitis by retrograde spread through vessels like anterior facial, angular and ophthalmic vein.

Clinical Features

- *Progress*—progress of orbital infection posteriorly may involve the superior orbital fissure and spread to cavernous sinus via superior ophthalmic vein (**Fig. 19-50**).
- *Symptoms*—it may result in temporary loss of visual acuity. Long-term ophthalmologic sequelae include permanent loss in visual acuity, residual proptosis, diplopia and blindness.
- *Signs*—swelling is seen in orbital region.
- *CNS involvement*—when later CNS is involved, hemiparesis, seizures and death have also been reported.



Fig. 19-50: Swelling seen in orbital region due to odontogenic infection of molar.

Diagnosis

- *Clinical diagnosis*—temporary loss of visual acuity with swelling in orbital region.

Management

- *Antibiotic*—when orbital infection is suspected, early aggressive antibiotic therapy and surgical intervention, following appropriate consultation, should be initiated to prevent serious ocular complications and CNS involvement.

Mediastinitis

It is usually a late complication of facial infection. Acute Mediastinitis when develops as complication of odontogenic infection, is called as *descending necrotizing mediastinitis (DNM)*. Such patient may have fulminating course and leading to death.

Pathogenesis

The process occurs because it gives a pathway for spread of pus and infection from submandibular region, floor of mouth and from all the related spaces in neck, beneath the investing layer to deep cervical fascia.

Following this pathway, infection can come into close relationship with trachea, larynx and great vessels and eventually reach the mediastinum.

The most common anatomic pathway is lateral pharyngeal space, through visceral space inferiorly, which may occur causing weakening and rupture of pleura.

Clinical Features

- *Symptoms*—patient complaint of face swelling, swallowing difficulty. Usually, there is chest pain and severe dyspnea with intermitting fever and evidence of swelling on the lateral aspect of neck of the affected side. Involvement of one or two facial spaces is usually preceded by these signs and symptoms.
- *Signs*—progressive septicemia, mediastinal abscess, pleural effusion, empyema, compression of mediastinum veins with decreased venous return to heart and pericarditis may occur, with death as the final step.
- *Hemorrhage*—hemorrhage secondary to erosion of internal carotid artery or one of its branches is the most common cause of death in deep space infection.

Radiological Features

- *CT scans*—this is more precise and accurate method than conventional radiography. On CT scan extent of necrosis can be determined.

Diagnostic Criteria for Diagnosis of DNM

- Clinical evidence of severe oropharyngeal infection.
- Characteristic radiological features of mediastinitis.
- Documentation of necrotizing mediastinitis infection at the operation or postmortem.
- Establishment of relationship between DNM and oropharyngeal process.

Management

- *Airway management*—utmost priority is to be given for airway control. In such cases, either emergency tracheostomy or cricothyroidectomy may be performed.

- **Antibiotics**—specific antibiotic therapy in high doses intensive antibiotic therapy instituted. Most commonly used antibiotics are MEPM which is carbapenem antibiotics. It should be used with cilastatin. Other antibiotics which can be used is clindamycin.
- **Surgical drainage**—the initial surgical drainage and debridement must be performed upto full extent.

Septicemia and Bacteremia

- **Bacteremia**—it refers to the circulation of bacteria in blood. Transient bacteremia is frequent even in healthy persons. In such cases, bacteria, are mopped up by circulating phagocytic cells and are unable to initiate infection.
- **Septicemia**—it implies overwhelming bacterial proliferation and release of toxins in blood. There is source of infection, hypotension and pyrexia with rigors.
- **Septic shock**—it refers to a condition of shock, secondary to sepsis. It is characterized by inadequate perfusion of tissue, which occurs due to septicemia with gram-negative infection. The inadequate perfusion of oxygen in the tissues is attributed to primary cellular defect in the utilization of oxygen, due to direct effect of sepsis. Despite the better understanding of this entity, newer treatment regimens and development of more potent antibiotics. Mortality rate remains above 50%. Majority of the septic processes that result in shock usually involve gram-negative bacteria. This shock state may also be caused by gram-positive bacteria.
- **Hyperdynamic shock**—hyperdynamic changes are altogether different from that of gram-negative sepsis and shock. Usually, there is hypotension, low peripheral resistance, peripheral vasodilation and normal cardiac output. This state of shock is characterized by dry warm skin. The so called *warm shock* is readily corrected by use of appropriate antibiotics and correction of fluid volume deficit. Patient response is favorable and survival rate is substantially high.
- **Gram-negative sepsis and shock**—this type of shock occurs when severe sepsis or endotoxemia is allowed to persist. The capillary membrane starts to leak and endotoxin is absorbed into blood, leading to generalized inflammatory state. Generalized capillary leakage and other fluid losses lead to severe hypovolemia, hypotension, reduce cardiac output, high peripheral resistance and peripheral vasoconstriction. A systemic infection results in cardiac depression, pulmonary hypertension, pulmonary edema and hypoxia, which in turn reduce the cardiac output. Progressive pulmonary insufficiency is characteristically seen. If infection is not controlled, there is rapid deterioration of pulmonary function, which leads to severe hypoxemia and death. The patient becomes cold, clammy, drowsy and tachypneic.

Necrotizing Fasciitis

This condition was first recognized in 1924 by Meleney. It is defined as rapidly progressing necrosis of subcutaneous tissue and fascia, usually sparing the muscles and accompanied by toxicity, high fever and apathy. Necrotizing Fasciitis is an inflammation of the connective tissue, which may be caused by streptococcal or other types of infection, an injury, or an autoimmune reaction.

Etiopathogenesis

- **Microorganism**—the usual cause is a mixture of aerobic and anaerobic organisms. Streptococcus A group is responsible.
- **Extension of organism to the subcutaneous tissue**—the organisms reach the subcutaneous tissue by extension from a contiguous infection or trauma to the area, including surgery.
- **Dermal gangrene**—there is widespread damage to the surrounding tissue, and occlusion of small subcutaneous vessels leads to dermal gangrene.
- **Risk factors**—the presence of diseases such as diabetes mellitus, malignancy and drug addiction, are significant risk factors.

Clinical Features

- **Symptoms**—the classic warning signals are unusually severe pain at the site of a wound or cut, or in the lymph nodes, and flu-like symptoms, surfacing a few hours after an injury or surgical operation.
- **Signs**—a tender erythematous cellulitis with ill defined margins develop. The affected area of skin becomes anesthetic, secondary to cutaneous nerve destruction, which can occur before clinical gangrene.
- **Progress**—the progression of disease can alarmingly rapid with skin color changing from red blue to grey in as early as 36 hours leading to frank cutaneous gangrene due to thrombosis of nutrient vessels, usually by 4th or 5th day.
- **Skin**—skin bullae may develop, but lymph adenitis is usually not seen. Skin death subsequent to subcutaneous necrosis is common (**Fig. 19-51**).

Diagnosis

This problem is diagnosed both clinically by an experienced clinician, based upon the rapid and severe progression of an infection, and by culturing the offending bacteria.

Management

- **Antibiotics**—spectrum of antibiotics should include drugs active against anaerobic organisms such as metronidazole.



Fig. 19-51: Necrotizing fasciitis showing destruction of subcutaneous tissue and exposing the bone.

- *Surgical debridement*—extensive surgical incision and debridement is the mainstay of treatment.
- *Hyperbaric oxygen therapy*—hyperbaric oxygen therapy has been used, but its value is not proven.
- *Supportive method*—blood transfusion and general supportive measures must be given.
- *Split skin graft*—as soon as disease is controlled split skin graft should be applied, if necessary for reconstruction.
- *Wound care*—continuous wound care is of utmost important. Wounds and cuts be well cleansed, kept scrupulously clean, disinfected. Minor wounds are best cleaned with soap and water, avoiding creams and ointments. Dressings put on wounds should be sterile and act as a filter—keeping out bacteria but allowing air to reach the wound. If sweat cannot evaporate and the cut stays moist, infection easily sets in, and bandaged. Irrigation with hydrogen peroxide, followed by dressing of charcoal lime and boric acid solution soaked gauze should be applied.

Pulpal Infection

Necrosis of Pulp

It is the death of pulp. It may be partial or total depending on whether a part or the entire pulp is involved.

Etiology

- *Trauma*—it is sequelae of inflammation and can also occur following trauma, in which the pulp is destroyed before an inflammatory reaction.

Types

- *Coagulation necrosis*—the soluble portion of tissue is precipitated or is converted into a solid material.

Caseation is a form of coagulation necrosis in which the tissue is converted into a cheesy mass consisting chiefly of coagulated proteins, fats, and water.

- *Liquefaction necrosis*—it results when proteolytic enzymes convert the tissue into softened mass, liquid or amorphous debris.

Clinical Features

- *Symptoms*—it does not cause painful symptoms.
- *Sign*—discoloration of the tooth is the first indication that the pulp is dead.
- *Thermal testing*—the tooth with partial necrosis can respond to thermal changes owing to presence of vital nerve fibers passing through the adjacent inflamed tissue.

Diagnosis

- *Clinical diagnosis*—history of severe pain lasting from a few minutes to a few hours, followed by complete and sudden cessation of pain.
- *Laboratory diagnosis*—there is necrotic pulp tissue, cellular debris and microorganisms may be seen in the pulp cavity. The periapical tissue may be normal or slight evidence of inflammation of the apical periodontal ligament may be seen.

Management

- *Root canal treatment*—preparation and obturation of root canals.

Pulp Calcifications

Various forms of pulp calcifications are found within the pulp which may be located in the pulp chamber or in the root canals. It can occur in any sex and in any teeth in the dental arch.

Etiology

- *Infection*—there is no clear-cut etiology. There is no relation between pulpal inflammation and irritation, as that arising from caries or trauma, since pulp calcification can be found in unerupted teeth. Extremely high percentage of pulp stones yield pure growth of streptococci on culture but often the affected teeth are normal.
- *Sundell schematic presentation*—local metabolic dysfunction → trauma → Hyalinization of injured cell → vascular damage → thrombosis vessel wall damage → fibrosis → mineralization (nidus formation) → growth → pulp stones.

Classification

- *Discrete pulp stones (denticles, pulp nodules)*—it is of following types:

- **True denticles**—they are made up of localized masses of calcified tissue that resembles dentin due to their tubular structure. Tubules are irregular and few in number. More common in pulp chamber than root canals. They are subdivided into:
 - *Free denticle*—denticle lying entirely within pulp tissue and is not attached to the dentinal wall.
 - *Attached denticle*—these are continuous with the dentinal wall.
- **False denticle**—it is composed of localized mass of calcified material and they do not exhibit dentinal tubules. Nodule appears to be made up of concentric layers or lamellae deposited around a central nidus. It is composed of cells around which laid down is a layer of reticular fibers that subsequently calcifies. They are again classified as free or attached. They are larger than true denticles and they may fill nearly the entire pulp chamber.
- **Interstitial denticle**—as the concentric deposition of calcified material continues, it approximates and finally is in apposition with the dentinal wall where it may be surrounded by secondary dentin, then is called as an interstitial denticle.
- **Diffuse calcification**—it is also called as 'calcific degeneration'. It is amorphous unorganized linear strands or columns parallel with blood vessels and nerves of pulp.

Clinical Significance

- **Symptoms**—sometimes, it may cause pain from mild pulpal neuralgia to severe excruciating pain resembling that of *tic douloureux* as the denticle can impinge on the nerve of the pulp.
- **Difficulty in root canal treatment**—difficulty may be encountered in extirpating the pulp during root canal therapy.

Radiographic Features

- **Appearance**—they are seen as radiopaque structures within the pulp chamber (**Fig. 19-52**), in the root canals or extending from pulp chamber into root canals.
- **Shape**—they may be round or oval. Their outline varies from sharply defined to more diffuse margins.
- **Location**—they may occur as single dense mass or several opacities.

Diagnosis

- **Clinical diagnosis**—it is not possible to make clinical diagnosis
- **Radiological diagnosis**—round or oval shaped radiopacity in the pulp chamber will diagnose this condition.

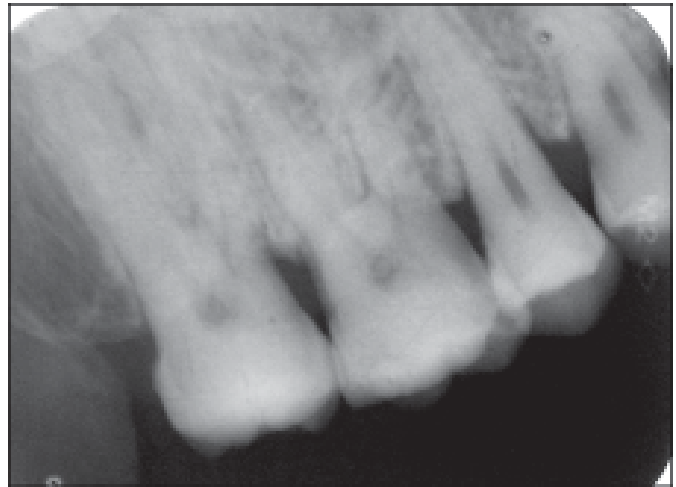


Fig. 19-52: Pulp stone seen as radiopacity in the pulp chamber.

Management

It is not required.

Focal Infection

Focus of infection refers to circumscribed area of tissue, which is infected with exogenous pathogenic microorganisms and which is usually located near a mucous or cutaneous surface. *Focal infection* refers to metastasis from the focus of infection, of organisms or their products that are capable of injuring tissue.

Mechanism of Focal Infection

- **Metastasis of microorganism**—it may be spread by hematogenous or lymphogenous route. They get localized in tissues. Certain organisms have a predilection for isolating themselves in specific sites of the body.
- **Toxin and toxin products**—it is spread by bloodstream or lymphatic channels, from focus to a distant site, where they may initiate hypersensitive reaction in the tissues. One example is scarlet fever, which is due to erythrocyte toxin liberated by the infective streptococci.

Oral Foci of Infections

- **Infected periapical lesion**—particularly, those of chronic nature an area usually surrounded by the fibrous capsule, which effectively walls off or separates the area of infection from the adjacent tissues but do not prevent the absorption of bacteria or toxins. Periapical granuloma has been described as a manifestation of vigorous body defense and repair reaction, while cysts merely a progressive form of granuloma. Abscess occurs when the reparative and defensive phase is minimum.

Majority of investigators indicate that an unusually high percentage of periapical granuloma are the biologically sterile and for this reason the possibility such lesions giving rise to focal infection is minimum.

- *Teeth with infected root canals*—these are potential sources of dissemination of microorganisms and toxins. Most commonly it shows occurrence of a hemolytic streptococcus; which is the most important in etiology of rheumatoid arthritis and rheumatic fever.
- *Periodontal disease*—it is equally significant as potential source of infection. The usual organism recovered is *Streptococcus viridans*. Simple massage of gingiva may result in transitory bacteremia. The rocking of teeth in their socket by forceps, before extraction, has been shown to favor bacteremia in patients who have periodontal disease. Due to pumping action during extraction microorganism may be forced from the gingival cervix into the capillary of gingiva as well as into the pulp of tooth and thus, will result in bacteremia. Oral prophylaxis may be followed by bacteremia. So it is mandatory to administer the antibiotics to the children who are diagnosed rheumatic or congenital heart disease; to prevent the positive consequences of bacterial endocarditis.

Significance of Oral Foci of Infection

There are reports that the oral foci of infection either cause, or aggravate many systemic disorders. Most common are as follows:

- *Arthritis*—it includes rheumatoid and rheumatic fever type. Arthritis of rheumatoid type is of unknown etiology. These patients have high antibody titer to group of hemolytic streptococci. It is tissue hypersensitive reaction.
- *Points in favor of septic foci theory*—certain point like streptococcal infection of throat, tonsils or nasal sinus may precede the initial or recurrent attack. Dramatic improvement occurs sometimes after the removal of septic foci. Temporary bacteremia may occur immediately after tonsillectomy, tooth extraction or after vigorous massage of gums.
- *Points against septic foci theory*—there is no infective focus can be found. Usually when focus is extirpated, no dramatic result is found. Sulfonamide, antibiotics and vaccine have failed to produce desirable results.
- *Valvular heart diseases*—subacute bacterial endocarditis is without doubt related to oral infections. There is close similarity, in most instances, between the etiologic agent of the disease and microorganisms in the oral cavity, dental pulp and in periapical lesions. Symptoms of subacute bacterial endocarditis have been observed in some instances shortly after extraction of teeth. Transient

bacteremia frequently follows tooth extraction. Streptococci of viridan type cause majority of subacute bacterial endocarditis. After tooth extraction, there is streptococcal bacteremia, so there is occurrence of subacute bacterial endocarditis after dental operations, dental extractions. Premedication of the patient should be done before extraction.

- *Gastrointestinal disease*—some workers state that constant swallowing of microorganisms might lead to variety of gastrointestinal diseases. Gastric and duodenal ulcers are produced by injection of streptococci.
- *Ocular disease*—factor supporting the hypothesis of Woods the role of foci of infection in ocular diseases.
 - *Ocular disease without systemic cause*—many ocular diseases occur in which no systemic cause, other than presence of remote foci of infection can be demonstrated.
 - *Healing of ocular disease after removal of oral foci*—numerous instances of prompt and dramatic healing of ocular diseases are reported following the removal of these foci.
 - *Exacerbation after removal of teeth*—occasionally, sudden transient exacerbation is observed, after the removal of teeth and tonsils.
 - *Blood stream infection*—presence of blood stream infection in early stages of ocular disease, are evident.
 - *Intravenous injection of microorganism*—iritis may be produced by intravenous injection of microorganisms, e.g. streptococci.
 - *Point against this theory*—many healthy people have focal infections, but do not have ocular diseases. Spontaneous cure may occur if nothing is done. Positive blood cultures are rare in acute iritis.
- *Skin diseases*—some forms of eczema and possibly urticaria, can be related to oral foci of infection. If the relationship does not exist, the mechanism is probably sensitization, rather than metastatic spread of the microorganisms.
- *Renal disease*—microorganism most commonly involved in urinary infection are *E.coli*, staphylococci and streptococci. Streptococci hemolyticus seems to be the most common. Streptococci are uncommon inhabitants of dental root canals or periapical and gingival areas. Since the microorganisms are commonly involved in renal infection, it appears that there is little relationship between oral foci of infection and renal disease.

Pits, Fistulae and Draining Sinus of Oral Cavity

- *Fovea palatini*—they are two indentations formed by coalescence of several mucus gland ducts near midline

of the palate. There is round to oval depressions always located in soft tissue on anterior part of soft palate. Depth of fovea is about 0.5 to 2 centimeters. It is accentuated when patient holds his nose and attempts to blow it. When manipulated they secrete 'clear mucinous fluid'.

- *Post-surgical pit*—it is a result of breakdown of the wound secondary to infection or failure to obliterate the dead space during wound closure. Dimple or puckering of either portion or entire surface of the wound with comparatively shallow depression is present. This can be probed easily.
- *Post-infection pit*—it results from loss of tissue often due to necrosis. After infection has been resolved, subsequent invasion of surface tissue into resultant defect forms post-infection pit. Surgical elimination.
- *Oronasal fistula*—it is a pathological defect lined by epithelium connecting the oral and nasal cavity. It may be congenital, palatal trauma, infection, neoplasm and surgical procedures. Sometimes acute dentoalveolar abscess burrow through maxilla in floor of nasal cavity and it may also be produced during surgical removal of teeth, buried roots, odontomas, tori, cysts and neoplasms. Patients complain of food passing into nose and often demonstrate nasal speech. Surgical removal of fistula and subsequent flap advancement.
- *Draining cyst*—odontogenic and non-odontogenic cysts may penetrate and produce a sinus that drains into the oral mucosa. Before sinus formation there is pain and swelling of the involved area. When periosteum and mucosa perforate, the pain ceases and purulent discharge is seen. If sinus is small, drainage may continue in chronic cases, but if it is large, cyst may disappear completely due to decompression. There is expansion of cortical plates.
- *Patent nasopalatine duct*—it is a rare condition; it arises when embryonic nasopalatine duct fails to obliterate. It is tunnel shaped and is continuous with the nasal epithelium. It extends downwards in an anterior direction more or less parallel with the facial contour or pre-maxilla to exist or seen on each side of palatine papilla. It is asymptomatic.
- *Pustule*—it is a small superficial elevation of skin and mucous membrane filled with pus and becomes a draining lesion for a short time after they rupture. They result from psoriasis, impetigo, acrodermatitis enteropathica containing superficial bacterial disease. It is surrounded by erythema. It is asymptomatic, but may be tender and painful.
- *Second branchial arch sinus*—it is fairly common and it occurs when the second branchial cleft or second pharyngeal pouch or both fail to obliterate during the embryological development of the fetus. It is having familial tendency. It can be unilateral or bilateral found

at birth or within the first year of life. It opens close to anterior border of sternocleidomastoid muscle with majority being in the lower neck above the sternoclavicular joint. There is history of intermittent drainage since childhood or spontaneous discharge by infection and cervical cyst. It appears as a small dimple or small opening in lateral region of neck.

- *Salivary gland fistula and sinus*—it is rare and is caused by accidental trauma, surgery and infection. It may occur as result of actinomycosis, syphilis and *carunculus oris*. The saliva escapes from damaged ducts either from pools within soft tissue or drains through fistula in skin.

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Introduction

Pigments are the colored substances, some of which are normal constituents of cells (e.g. melanin), whereas others are abnormal and collect in cells only after under special circumstances. Pigments can be *exogenous*, coming from outside in the body, or *endogenous*, synthesized within the body itself. Exogenous pigmentation is commonly due to foreign-body implantation in the oral mucosa.

Endogenous pigments include *melanin*, *hemoglobin*, *hemosiderin* and *carotene*.

In the course of disease, the oral mucosal tissues can assume a variety of discolorations blue, brown, and black discolorations constitute the pigmented lesions of the oral mucosa, and such color changes can be attributed to the deposition of either endogenous or exogenous pigments.

The most important endogenous factor causing pigmentation in the oral cavity is *melanin*. It is derived from the Greek (Melas-black), is an endogenous, non-hemoglobin derived, brown-black pigment formed when the enzyme tyrosinase catalyzes the oxidation of tyrosine to dihydroxy-phenylalanine in melanocytes. Melanin in the melanosome, the specialized epidermal melanin-bearing organelle, is responsible for the color variation of human skin.

There are generally considered to be two types of melanin pigmentation which form normal skin color. *Constitutive skin color* implies the genetically determined color of healthy unexposed skin such as observed on habitually sun-shielded areas like the buttock and inner upper arm; *facultative skin color* applies, for example, to “tanned” skin and reflects the genetic capacity of the skin to darken in response to ultraviolet radiation exposure.

Three types of melanin have been demonstrated.

- *Eumelanin*—it is brown-black melanin which is found in ellipsoid melanosomes which impart brown-black color to skin and hair.
- *Pheomelanin*—it is yellow-red melanin in spherical melanosomes which are the basis of yellow-red hair.

- *Neuromelanin*—it is black pigment formed within nerve cell by an enzymic pathway different from that responsible for eumelanin or pheomelanin formation.

The melanin producing cells, melanocytes, are highly specialized dendritic cells of neural crest origin. Melanocytes are found in humans in skin, mucous membrane, uveal tract. Melanin pigment synthesized in specialized cytoplasmic organelles called *melanosomes*, which contain the aerobic oxidase, tyrosinase, which initiates the conversion of tyrosine to melanin.

The endogenous pigmentation of the oral mucous membrane is most frequently explained by the presence of hemoglobin, hemosiderin, and melanin. Hemoglobin imparts a red or blue appearance to the mucosa and represents pigmentation associated with vascular lesions; the coloration is rendered by circulating erythrocytes coursing through patent vessels. In contrast, hemosiderin appears brown and is deposited as a consequence of blood extravasation, which may occur as a consequence of trauma or a defect in hemostatic mechanisms. Hemochromatosis (generalized hemosiderin tissue deposition) may occur as a result of a variety of pathologic states.

Essence of Oral Pigmentation

Pigmentation of the perioral and intraoral tissues is a frequent finding. Oral pigmentation may be physiologic or pathologic and the finding of oral pigmentation may represent a systemic disease of the patient. Such lesions represent a variety of clinical entities, ranging from physiologic changes (e.g. racial pigmentation) to manifestations of systemic illnesses (e.g. Addison’s disease) and malignant neoplasms (e.g. melanoma and Kaposi’s sarcoma). Finding of oral pigmentation may represent a systemic disease of the patient. Therefore, an understanding of the causes of mucosal pigmentation and appropriate evaluation of the patient are essential.

Gingival physiologic pigmentation may cause problems for some patients especially female as far as esthetics is concerned. Social behavior can be devastatingly affected by pigmentation disorder. The psychological problems arising from the pronounced cosmetic disfigurement of vitiligo patient in pigmented races are easily understood.

Definition and Terminologies Used in Pigmentation

- *Hypomelanosis*—it refers to decrease in normal melanin pigmentation.
- *Amelanosis*—It refers to total lack of melanin.
- *Depigmentation*—It refers to loss of previously existing melanin.
- *Leukoderma*—it is a generic term for skin relatively or absolutely lightened in color.
- *Poliosis*—it refers to localized whitening of hair.
- *Canities*—it refers to more generalized defect in whitening of hair.
- *Macule*—well-circumscribed, flat lesions that are noticeable because of their change from normal skin color. They may be red due to the presence of vascular lesions or inflammation, or pigmented due to presence of melanin, hemosiderin, and drugs.
- *Papules*—solid lesions raised above the skin surface that are smaller than 1 cm in diameter. Papules may be seen in a wide variety of diseases including erythema multiforme simplex, rubella, lupus erythematosus, and sarcoidosis.
- *Plaques*—solid raised lesions that are over 1 cm in diameter; they are large papules.
- *Wheal*—itchy, transient, elevated lesion with variable blanching and erythema formed as the result of dermal edema.
- *Hematoma*—it is localized collection of blood, usually clotted, in a tissue or organ.
- *Purpura*—reddish to purple flat lesions caused by blood from vessels leaking into the subcutaneous tissue. It is classified by size as petechiae or ecchymoses, these lesions do not blanch when pressed.
- *Petechiae*—purpuric lesions 1 to 2 mm in diameter.
- *Ecchymoses*—it is greater than 3 mm.

- *Burrow*—a linear or curvilinear papule, caused by a burrowing scabies mite.
- *Comedo*—a plug of keratin and sebum wedged in a dilated pilosebaceous orifice.
- *Telangiectasia*—the visible dilatation of small cutaneous blood vessels.
- *Scale*—a flake arising from the horny layer.
- *Crust*—looks like scale, but is composed of dried blood or tissue fluid.
- *Atrophy*—thinning of skin due to diminution of the epidermis, dermis, subcutaneous fat.
- *Stria*—a streak-like, linear, atrophic, pink, purple or white lesion of the skin due to changes in the connective tissue.

Classification

It is discussed in **Tables 20-1 to 20-3**.

Blue/Purple Vascular Lesions

Hemangioma

Hemangioma is a benign proliferation of the endothelial cells that line vascular channels. Vascular malformation is a structural anomaly of blood vessels without endothelial proliferation. Both lesions are developmental abnormalities, characterized by onset during infancy. Hemangioma regresses as the patient ages, but vascular malformation persists throughout life. Hemangioma of the head and neck region is relatively common, representing at least a third of all hemangiomas in humans. Oral hemangioma represents 14% of all human hemangiomas.

A simple classification is that proposed by Watson and McCarthy is as follow capillary hemangioma, cavernous hemangioma, angioblastic or hypertrophic hemangioma, racemose hemangioma, diffuse systemic hemangioma, metastasizing hemangioma, nevus vinosus or port-wine stain and hereditary hemorrhagic telangiectases.

Etiology

It is often congenital in nature and usually but not invariably follows a benign course. Some authorities believe

Table 20-1: First classification according to color

Color	Focal	Diffuse	Multifocal
Blue/purple	Varix, Hemangioma	Hemangioma	Kaposi's sarcoma, Hereditary hemorrhagic telangiectasia
Brown	Melanotic macules, nevus, melanoma	Ecchymosis, melanoma, drug induced, hairy tongue, petechiae	Physiologic pigmentation, neurofibromatosis, lichen planus, Addison's disease, drug induced, Peutz-Jegher's syndrome
Gray black	Amalgam tattoo, graphite tattoo, nevus, melanoma	Amalgam tattoo, hairy tongue	Metal ingestion

Table 20-2: Second classification

Diffuse and bilateral	Focal
• Early onset • Physiologic pigmentation • Peutz-Jegher's syndrome • Predominantly adults onset <i>With systemic signs and symptoms</i> • Addison's disease • Kaposi's sarcoma • Heavy metal pigmentation <i>No systemic signs and symptoms</i> • Smoker's melanosis • Drug-induced pigmentation • Post inflammatory pigmentation	• Red blue purple <i>Blanching</i> • Hemangioma • Varix <i>Non-Blanching</i> • Thrombus • Hematoma • Blue gray • Amalgam tattoo • Other foreign body tattoos • Blue nevus • Brown • Oral melanotic macule • Pigmented nevus • Melanoacanthoma • Melanoma

Table 20-3: Endogenous pigmentation

It is most often explained by pigments like hemoglobin, hemosiderin and melanin.		
Pigment	Color	Disease process
Hemoglobin	Blue, red, purple	Varix, hemangioma, Kaposi's sarcoma, angiosarcoma, hereditary hemorrhage telangiectasia
Hemosiderin	Brown	Ecchymosis, petechiae, varix, hemorrhage mucocele, hemochromatosis
Melanin	Brown, black, gray	Melanotic macule, nevus melanoma, basilar melanoma

the lesion, particularly the congenital form, is not the true neoplasm, but rather a developmental anomaly or hamartoma.

Clinical Features

- **Signs**—the earliest sign of a hemangioma is blanching of the involved skin, often followed by fine telangiectases and then a red macule.
- **Hemangioma in children**—rapid growth during the neonatal period is the hallmark of hemangiomas, occurring characteristically beyond the growth rate of the infant. The hemangiomas of childhood are found on the skin, in the scalp, and within the connective tissue of mucous membranes. Approximately 85% of childhood-onset hemangiomas spontaneously regress after puberty.
- **Diascopy**—diascopy usually shows blanching on pressure. This procedure is performed by pressing gently on the lesion with a glass slide or a glass test tube. A positive diascopy result (blanching) generally indicates that the blood is within vascular spaces and is displaced out of the lesion by pressure. However, lack of blanching does not exclude the possibility of a vascular lesion.

- **Color of oral hemangioma**—hemangioma of the oral soft tissue is similar to the hemangioma of the skin and appears as a flat or raised lesion of the mucosa. It is usually deep red or bluish red and seldom well circumscribed (**Fig. 20-1**).
- **Site**—the most common site of occurrence is the lips, tongue, buccal mucosa and palate. The tumor often is traumatized and undergoes ulceration and secondary infection.



Fig. 20-1: Bluish color pigmentation seen in case of hemangioma of tongue (Courtesy Dr Tapasya Karamore).

Differential Diagnosis

- **Mucocele, ranula and superficial cyst**—the hemangioma blanches and may be emptied by the application of digital pressure, which forces the blood from the vascular spaces. This feature accounts for the finding that the lesion is not fluctuant and, in turn, helps to differentiate the cavernous hemangioma from the mucocele, ranula and superficial cyst, which though soft are, in contrast, fluctuant and nonemptiable.
- **Varicosity**—a varicosity is usually seen as an elongated enlargement of superficial vein rather than as a nodule or dome shaped mass. Furthermore, a pulse is not detectable within the cavernous hemangioma.
- **Arteriovenous shunt or aneurysm**—no pulse detected in hemangioma as in case of arteriovenous shunt or an aneurysm, both of which may occur as rubbery, nonfluctuant, domelike, bluish nodules with a usually discernible throbbing.

Management

- **Sclerosing technique**—sclerosing agents such as sodium tetradecyl sulfate can be used intralesionally.
- **Cryosurgery**—it can be useful in treating hemangioma.

Varix and Varices

Varix is an enlarged and convoluted vein, artery or lymphatic vessel. It is an acquired benign vascular lesion, generally asymptomatic, and does not require treatment. Oral varix is morphologically composed by one to three extensive and tortuous blood vessels lined by flat mature endothelium.

Definition

- *Varices*—pathological dilatation of vein or venules are varices or varicosity.
- *Varix*—focal dilatation of group of venules or vein is known as varix.

Etiology

Trauma plays an important role in the development of a varix. The traumatic event probably damages and weakens the vascular wall and result in dilation.

Clinical Features

- *Location*—it is commonly found on the tongue, lip or cheek, mainly in the seventh decade of life.
- *Color*—oral varix is characterized as a red to purple papule or nodule.
- *Margins and shape*—it has sharply delineated borders and a smooth, rounded surface contour.
- *Caviar tongue*—when many of sublingual veins are involved it is called caviar tongue (**Fig. 20-2**).



Fig. 20-2: Caviar tongue appears showing many purple colored dilation of venules.

Differential Diagnosis

- *Difference between hemangioma and varix*—the varix resembles the hemangioma both clinically and histologically, yet it is distinguished by two features: the patient's age at its onset and its etiology. A hemangioma

is usually congenital and has a tendency to spontaneously regress whereas a varix arises in older individuals and, once formed, does not regress. Alternatively, a varix has a finite growth potential; once a varix has formed, further enlargement is uncommon. Whereas hemangiomas are vascular hamartomas of unknown etiology, the varix represents a venous dilatation that may evolve from trauma such as lip or cheek biting.

- *Superficial non-keratotic cyst and ranula*—it cannot be emptied by digital pressure.
- *Aneurysm*—rare and demonstrate pulse.
- *Hereditary hemorrhagic telangiectases*—it may confuse with varix but is multifocal and hemorrhagic.
- *Nevi*—nevi does not blanch on pressure and it has palpable nature.

Management

- *Surgical approach*—the lesion can be excised or removed by other surgical methods, including electrosurgery and cryosurgery.
- *Intralesional injection of sclerosing solution*—intralesional 1% sodium tetradecyl sulfate injection is effective as well, yet it is usually more painful than simple excision. This sclerosing agent should be injected directly into the lumina with a tuberculin syringe (depositing 0.05 to 0.15 mL/cm³).

Thrombus

A thrombus, or blood clot, is the final product of the blood coagulation step in hemostasis. It is achieved via the aggregation of platelets that form a platelet plug, and the activation of the humoral coagulation system (i.e. clotting factors). A thrombus is physiologic in cases of injury, but pathologic in case of thrombosis.

Specifically, a thrombus is a blood clot in an intact blood vessel. A thrombus in a large blood vessel will decrease blood flow through that vessel. In a small blood vessel, blood flow may be completely cut-off resulting in death of tissue supplied by that vessel. If a thrombus dislodges and becomes free-floating, it is an embolus.

Etiology

- *Damage to vessel*—both arterial and venous thrombosis may arise either because of damage to the vessel for instance, atheroma or varicose veins.
- *Change cellular elements*—it may result due to changes in the plasma or cellular elements.

Clinical Features

- *Color*—if the varix contain the thrombus, it presents as a firm bluish purple nodule that does not blanch under pressure.

- *Site*—thrombi are more common on the lower lip and buccal mucosa.

Management

- *Heparin and warfarin*—these are often used to inhibit the formation and growth of existing blood clots, thereby allowing the body to shrink and dissolve the blood clots through normal method.

Kaposi's Sarcoma

Synonyms: Multiple idiopathic hemorrhagic sarcoma of Kaposi's, Angioreticuloendothelioma.

Kaposi's sarcoma is an unusual and uncommon disease of blood vessels which occasionally manifests in the oral cavity: Classical (sporadic) Kaposi's sarcoma was described by Moritz Kaposi in 1872 in central Europe among elderly persons of Mediterranean or Jewish origin as a multiple pigmented sarcoma of the skin. Kaposi's is most accurately described as a multifocal angioproliferative neoplasm that primarily involves the skin in non-HIV infected person. KS is a neoplasm now most commonly a manifestation of infection with the human immunodeficiency virus (HIV), that frequently affects the oral cavity.

Although KS interaction with HIV infection remains unexplained, the epidemic occurrence of KS in patient with AIDS clearly supports a causal relationship between the immune deficiency and the appearance of this neoplasm. Pathogens or factors suspected to be associated with KS include human herpes virus type 6, cytomegalovirus, human immunodeficiency virus, mycoplasma penetons, sex hormones and nitrate inhalants. However, solid evidence of etiologic association among any of these agents with AIDS or non-AIDS KS has not been established yet.

This vascular neoplasm may present as cutaneous, oral or visceral lesion either independently or synchronously. The frequency is higher in homosexual and bisexual male than in other risk group for HIV infection: There is also an increased incidence of Kaposi's sarcoma among transplant patients, which can resolve after immunosuppression drug therapy is halted. Some investigators believe that KS is a form of vascular hyperplasia responding to endothelial growth factors rather than a malignant neoplasm.

Clinical Features

- *Age distribution and sex distribution*—KS may occur at any age, but is most common in the fifth, sixth and seventh decades. There is marked male predilection with a gender ratio of 20 to 1.
- *Appearance*—the tumor begins as a multicentric neoplastic process that manifests as multiple red/purple (vascular appearing) macules and in more advanced types, nodules, occurring on the skin or mucosal areas.
- *Progress*—the lesion tends to enlarge and become darker and may coalesce or form clusters of single nodules.
- *AIDS associated*—occurrence of KS in patient with AIDS in the head-neck area is common. The tip of the nose is a peculiar but frequent location of these. Facial, scalp, periorbital and conjunctival involvement are also typical. The neoplasm can involve lymph nodes, soft tissue of the extremities and GIT.
- *Clinical presentation*—four clinical presentation of Kaposi's sarcoma are recognized.
 - *Classic type*—classic (chronic) KS is primarily a disease of late adult life and about 90% of cases occur in man. It mostly affects individuals of Italian, Jewish or salvic ancestry. Multiple bluish purple macules and plaques are present on the skin of the lower extremities.
 - *Endemic*—endemic KS in Africa has been divided into four subtypes:
 - *A benign nodular type* similar to classic KS.
 - *An aggressive type*, is characterized by progressive development of locally invasive lesions that involve the underlying soft tissue and bone.
 - *A florid form* is characterized by rapidly progressive and widely disseminated, aggressive lesions with frequent visceral involvement.
 - *A unique lymphadenopathic type*, which occurs primarily in young black children and exhibits, generalized, rapidly growing tumors of lymph nodes, occasional visceral organ lesions and spare skin involvement.
 - *Iatrogenic type*—iatrogenic immunosuppression associated KS most often occurs in recipient's organ transplants. It affects 0.4% of renal transplant patient, usually several months to a few years after the transplant. It is probably related to the loss of cellular immunity, which occurs as a result of immunosuppressive drug.
 - *AIDS related KS*—frequently involves the head and neck.

Oral Manifestation

- *Location*—the lesions of the oral mucosa are identical in appearance with cutaneous nodules. KS can involve any oral site but most frequently involves the attached mucosa of the palate, gingiva and dorsum of tongue.
- *Appearance*—oral KS lesions occur as red to purple nodules and macule with mucosal ulceration in some of the more mature cases.
- *Symptoms*—the lesions result in pain, dysphagia, difficulty with mastication, bleeding, and may be cosmetically displeasing. This is particularly true with the progression from the flat to the nodular form, which has been associated with increasing grades of immunocompression.

- *Palpation*—the lesion does not blanch with pressure. Early lesion may appear as similar to ecchymosis or hemangioma.

Differential Diagnosis

- *Hemangioma*—it blanches on pressure.
- *Purpura*—it is presented as multiple papules on the soft palate.
- *Nevi*—it is not so aggressive lesion.
- *Melanoma*—it is not multicentric process.

Management

- *Radiation therapy*—for skin lesions in the classic form of the disease, radiation therapy often is used. Radiotherapy for oral KS is most often fractionated to result in a dose of 25 to 30 Gy over 1 week. Severe mucositis can follow radiotherapy for oral KS, although this may be less severe with fractionated treatment.
- *Surgical eradication*—surgical eradication of the disease is difficult because of the multiplicity of lesions. The carbon dioxide or argon laser can be very effective in the surgical treatment or palliation of KS involving the upper aerodigestive tract.
- *Systemic chemotherapy*—systemic chemotherapy especially vinblastine may also be helpful. If KS progresses at multiple sites, systemic chemotherapy may be needed. The most commonly used regimen is alternating weekly vinblastine and vincristine. Other effective agents include doxorubicin in low doses, bleomycin and methotrexate. Etoposide has also been used. Recombinant interferon alpha—2A and alpha—2b have been identified as efficacious agents in the treatment of KS.
- *Intralesional injection*—the intralesional use of vinblastine, in doses varying from 0.01 to 0.04mg (multiple intralesional injections), may be an effective alternative treatment for small KS lesion of the mouth. The treatment is repeated every 2-4 weeks until the remission of the tumor mass is obtained.
- *Combination therapy for AIDS related Kaposi's sarcoma*—in case of epidemic (AIDS associated) KS, a great variety of treatment regimens such as single agents, combined chemotherapy, or zidovudine with interferon—alpha and radiotherapy have been tried. Circumscribed lesions localized within the mouth and associated with a CD4 T-lymphocyte count of more than 500 cells /mL may be treated by excision, cryotherapy, or even sclerotherapy, to which they rapidly respond. At a CD4 level of fewer than 200 cells/mL, systemic chemotherapy and anti-retroviral treatment is advisable; while for intermediate CD4 counts, interferon with zidovudine may be appropriate. Prognosis is variable, depending on the form of disease and the patient's immune status.

Hereditary Hemorrhagic Telangiectasia

- *Cause*—it is a genetically transmitted disease inherited as an autosomal dominant trait.
- *Site*—there may be hundreds of such purple papules on the vermillion border and mucosal surface of lips as well as tongue and buccal mucosa.
- *Color*—papules are red or brown rather than purple.
- *Shape and size*—it is characterized by multiple, round or oval papules measuring less than 0.5 cm in diameter.
- *Management*—it can be cauterized by electrocautery.

Blue Nevus

Nevus is defined as a congenital, developmental tumor-like malformation of the skin or mucous membrane. It is a true mesodermal structure composed of dermal melanocytes which only rarely undergo malignant transformation.

The Blue nevus represents a localized proliferation of dermal melanocytes. It is an uncommon pigmented tumor of dermal melanocytes that has traditionally been classified into common and cellular variant. It is usually a skin tumor in adults but can become apparent in early childhood or even present at birth. Malignant blue nevus is a rare melanocytic tumor of the skin arising from a preexisting cellular blue nevus.

Etiology

- *Dermal arrest*—although definitive experimental evidence is lacking, blue nevi are believed to represent dermal arrest in embryonal migration of neural crest melanocytes that fail to reach the epidermis.
- *Genetic predisposition*—because of the variation of blue nevi in different populations, a genetic predisposition has been suggested. However, familial cases of blue nevi are exceedingly rare.

Clinical Features

- *Age and sex predilection*—blue nevi are twice more common in women than in men. Blue nevi may develop at any age but are usually noticed in the second decade of life or later.
- *Site*—it occurs chiefly on buttocks, on the dorsum of feet and hands, on the face and occasionally on other area.
- *Color*—the lesion is smooth, exhibits hairs growing from its surface and varies in colors from brown to blue or bluish black.
- *Appearance*—the common blue nevus is a flat to slightly elevated, smooth surfaced macule, papule, or plaque that is gray-blue to bluish black in color.
- *Tyndall effect*—the clinically noted blue color is due to the depth of melanin in the epidermis and has the Tyndall effect. The Tyndall effect is the preferential

absorption of long wavelengths of light by melanin and the scattering of shorter wavelengths, representing the blue end of the spectrum, by collagen bundles.

- *Cellular blue nevus*—the cellular blue nevus was first described as a variant of melanoma. Later, it was classified as a variant of blue nevus. Controversy still arises over the precise distinction of atypical cellular blue nevus from melanoma. The cellular blue nevus is a less common lesion but often clinically similar to the common blue nevus. These lesions tend to be large, usually measuring 1-3 cm in diameter. Lesions are elevated, smooth-surfaced papules or plaques that are gray-blue to bluish black in color. Lesions are usually solitary and found on the buttocks, the sacral region, and occasionally on the dorsal aspects of the hands and the feet.

Differential Diagnosis

- The differential diagnosis includes traumatic tattoo, pigmented basal cell carcinoma and melanoma.

Management

- *Surgical excision*—simple excision is the treatment of choice.

Brown Melanotic Lesions

Melanotic Macule

The term 'melanotic macule' has been used to describe a benign pigmented lesion of the oral cavity, characterized by an increase in melanin pigmentation along the basal cell layer of the epithelium and the lamina propria. Melanotic macules have been variously termed as *ephelis*, *melanosis*, *lentigo*, *solitary labial lentigo*, *labial melanotic macule* and *oral melanotic macules*. It represents an increase in synthesis of melanin pigments by basal cell layer melanocytes without increase in the number of melanocytes. It is the most common pigmentation to occur in oral cavity of light skinned individuals.

Etiology

- *Genetic*—controversy exists on the pathogenetic mechanism that leads to the development of melanotic macules and it is not clear if they represent a physiologic or a reactive process. Some authors reported a positive family history and a genetic predisposition has been hypothesized.
- *Racial*—if arising in children, it is most likely racial in origin, in which case it may be called racial pigmentation (Fig. 20-3) or physiologic pigmentation; no treatment is necessary.

- *Environmental factors*—when it arises in adults it may be smoke-induced, drug-induced, hormone-induced or spontaneous (without cause), and it is usually biopsied in order to be sure that it is not a malignant melanoma. Some inherited diseases show brown pigmentation of the oral membranes.



Fig. 20-3: Racial pigmentation seen on the gingiva.

Clinical Features

- *Age*—The solitary oral melanotic macule is seen in middle aged adults. Females are more affected than males.
- *Site*—it is attributed to actinic exposure and therefore occurs on vermilion border of the lower lip. Sometimes it can also occur on gingiva, palate and buccal mucosa.
- *Color and appearance*—the melanotic macule is typically a well circumscribed flat area of pigmentation that may be brown, black, blue or gray in color.
- *Size*—most of the lesions are less than 1 cm in diameter, although in occasional cases, they may be larger in size.
- *Shape*—lesions are oval or irregular in outline.

Differential Diagnosis

- *Melanoplakia*—they are larger and occur in black individuals.
- *Amalgam tattoo*—associated with juxtaposed amalgam filling.
- *Ecchymosis patch*—it has got brownish color and it usually disappears within few days.
- *Superficial spreading melanoma*—It is seen in older age group and spreads by circumferential growth. Men are more affected and palate is commonly affected site.
- *Flat nevi and lentigo*—are very rare in the oral cavity.
- *Focal melanosis*—it is not seen as pigmented lesion.

Management

- *Surgical excision*—excision with adequate borders should be carried out.

Pigmented Nevus

Nevus is the general term that may refer to any congenital lesion of various cell types or tissue types, such as epidermis, vessels, and pigment cells. It is a congenital or acquired benign tumor of melanocytes or nevus cell that occurs on skin. Oral nevi are rare. It is usually but not always pigmented, ranging from gray to light brown to blue to black. They are of four types—intramucosal, junctional, compound and blue nevus.

Etiology

- *Pigment cells*—the origin of nevus cells is not completely known. It has been postulated that they are derived from pigment cells that migrate from the neural crest to the epithelium and dermis (submucosa) or that may develop from altered resident melanocytes.

Clinical Features

- *Age*—they usually appear shortly after birth and throughout childhood.
- *Site*—it is most frequently encountered on the palate, gingiva, buccal mucosa and lip.
- *Color and appearance*—intraoral nevi are rare that may occur at any age. It is characterized by a plaque or dome-shaped sessile nodule with blue or black pigmentation (Fig. 20-4).
- *Signs*—pigmented nevi do not blanch on pressure. They generally reach a given size and then growth becomes static.



Fig. 20-4: Pigmented nevus seen on neck of patient

Differential Diagnosis

- *Vascular lesions*—these include hematoma, varix, and hemangioma. Hematoma is ruled out by history and varix and hemangioma are ruled out by diascopy (compression) method. These vascular lesions blanch on pressure.

- *Amalgam tattoo*—amalgam restoration is seen adjacent to pigmentation.
- *Melanoma*—they are rare in the oral cavity.

Management

- *Excisional biopsy*—junctional nevi in adult may evolve into malignant melanoma. For this reason, all nevi should be excised. Since their size is generally less than 1 cm, excisional biopsy would be usually indicated.

Melanoacanthoma

The term 'melanoacanthoma' was first used by Mishima and Pinkus in 1960 to describe a benign mixed skin tumor composed of basal and prickle cell keratinocytes and pigment-laden dendritic melanocytes. Oral melanoacanthoma is a rare, benign pigmented lesion, similar to cutaneous melanoacanthoma, characterized by proliferation of dendritic melanocytes scattered throughout the thickness of an acanthotic and hyperkeratotic surface epithelium.

Etiology

- The pathogenesis of oral melanoacanthoma is not clear, although its clinical behavior is suggestive of a reactive origin.

Clinical Features

- *Sex predilection*—the lesion occurs almost exclusively in blacks and has a female predilection.
- *Site*—it is seen most frequently on the buccal mucosa, which may be related to greater frequency of trauma in this area in 20-40-year-old persons. But can occur at any oral site like lip, palate and gingiva and at any age.
- *Appearance*—it is usually presenting as a single smooth, flat or slightly raised, dark brown to black macule.
- *Size*—the lesion often demonstrates an alarmingly rapid increase in size and occasionally will reach a diameter of several centimeters within a period of a few weeks.
- *Symptoms*—it is asymptomatic and not indurated.

Differential Diagnosis

- *Melanoma*—this lesion has a tendency to enlarge rapidly, which raises the possibility of a malignant process in the clinical differential diagnosis. However, its tendency to occur in young black females distinguishes it from melanoma, which is uncommon in this age and racial group.

Management

- *Incisional biopsy*—oral melanoacanthoma appears to be a reactive lesion with no malignant potential. In some cases, the lesion disappears after incisional biopsy or removal of the offending stimulus.

Melanoplakia

This term is applied to a flat, localized or widespread, black or brownish discoloration of the oral mucosa due to increased amount of melanin.

Melanin is produced by dentrite melanocytes in basal cell layer of epidermis and is formed by oxidation of tyrosine, a reaction that is catalyzed by copper containing enzyme tyrosinase and mediated by melanocytes stimulating hormone from anterior pituitary.

Etiology

- *Smoker melanosis*—it may occur due to smoker's melanosis.
- *Racial pigmentation*—it may occur due to racial pigmentation.
- *Systemic disorders*—it can also occur in diseases like Peutz-Jegher's syndrome, Addison's disease or hemochromatosis or may result from lead poisoning or cancer chemotherapy.

Clinical Features

- *Race*—dark complexioned people frequently have macular pigmentation of various sizes on their oral mucosa.
- *Color*—it varies from light brown (Fig. 20-5) to blue pigmentation depending on amount of melanin present and depth of lesion.



Fig. 20-5: Melanoplakia showing light brown type of pigmentation.

Differential Diagnosis

- *Amalgam tattoo*—tooth with amalgam filling is seen in close proximity.
- *Junctional nevus*—it is very rare in oral cavity.
- *Melanoma*—it is seen in older individuals and spread by circumferential growth.
- *Local area of hemosiderin deposit* —it is not seen as pigmented lesion.

Management

- *Biopsy*—biopsy is done if the pigmented patch appears recently rather than from birth or during childhood. It becomes elevated in part or whole lesion and increases in size and undergoes color changes and surface ulceration.

Smoker's Melanosis

Smoker's melanosis occurs in 25 to 31% of tobacco users and is characterized by discrete or coalescing multiple brown macules that usually involve the attached mandibular gingiva on the labial side, although pigmentation of the palate and buccal mucosa has also been associated with pipe smoking. Smoking-associated melanosis is due to increased melanin production by melanocytes and its deposition within the basal cell layer and lamina propria. The microscopic appearance of melanosis is essentially similar to that seen in physiologic pigmentation or a melanotic macule.

Etiology

- *Nicotine*—smoker's melanosis may be due to the effects of nicotine (a polycyclic compound) on melanocytes located along the basal cells of the lining epithelium of the oral mucosa (Fig. 20-6). Nicotine appears to directly



Fig. 20-6: Smoker's melanosis presented as black type of pigmentation.

stimulate melanocytes to produce more melanosomes, which results in increased deposition of melanin pigment as basilar melanosis with varying amounts of melanin incontinence.

- *Polycyclic amines*—Araki et al reported that polycyclic amines in tobacco, such as nicotine and benzpyrenes, stimulate melanocytes to increase their melanin production. It has been suggested that oral melanin may protect the mucosa by binding with toxic agents from tobacco smoke, which can readily penetrate into the tissue. Smoker's melanosis seems to be directly related to stimulant effect of substances in smoke on melanocytes.

Clinical Features

- *Race and sex distribution*—this condition is most evident in whites because of a lack of physiologic pigmentation in the oral mucosa of this population, but some dark-skinned individuals who smoke will have more prominent pigmentation in many oral sites. Females are affected more than males, which may be explained by the additive effects of estrogen in female smokers. Increases in estrogen levels observed during pregnancy and the use of birth control pills are linked to other hyperpigmentation conditions (e.g. melasma).
- *Age*—the incidence of smoker's melanosis increases with age, suggesting that the longer a person smokes, the more likely he or she will develop the condition. Some authors noticed a significant increase in gingival melanosis of children with parents who smoke.
- *Location*—smoker's melanosis has been mostly observed at the anterior mandibular gingiva; however, it can also be seen in other parts of the oral cavity. Other part involved is palate (Fig. 20-7), buccal and commissural mucosa, and inferior surface of tongue and lip mucosa.



Fig. 20-7: Heavy pigmentation seen in smoker melanosis.

- *Color*—smoker's melanosis has usually black-brown pigmentation. The prevalence of clinically visible oral melanin pigmentation is directly correlated with the number of cigarettes consumed per day. The consumption of 1-3 cigarettes/day elevates the prevalence of oral pigmented lesions from 3% among nonsmokers to 9%. When patients smoke more than 15 cigarettes/day, the prevalence increases to 31%.

Management

With cessation of the smoking, improvement is expected over the course of months to year. Smoker's melanosis, appears to be little significance. It may, however, potentially mask other lesions or may be cosmetically objectionable.

Differential Diagnosis

Other entities to consider in differential diagnosis are racial melanosis, melanosis due to medications, Peutz-Jegher's syndrome, Addison's disease and early melanoma. Smoker's melanosis is benign and not considered to be precancerous, but a biopsy may be indicated to rule out more serious conditions, in particular melanoma.

Melanoma

It is also called as *melanocarcinoma*. Melanoma is the malignant neoplasm of melanocytic origin that arises from a benign melanocytic lesion or *de novo* from melanocytes within otherwise normal skin or mucosa. It is the third most common cancer of skin but they may develop at any site where melanocytes is present.

Primary malignant melanoma of the oral mucosa is extremely rare tumor, representing 0.2 to 8% of all melanomas. Amelanotic malignant melanoma accounts for only 2.3% of all melanomas. Amelanotic malignant melanoma was defined as a tumor composed of nonpigmented melanocytes. However, another report, a tumor that lacks pigmentation clinically and has melanin pigmentation histopathologically is also included in this category. It is neoplasm of epidermal melanocytes.

Growth Phase

- *Radial growth phase*—radial growth phase is the initial phase of growth of the tumor. During this period, which may last many years, the neoplastic process is confined to the epidermis. In the early stages of melanoma development, the radial growth phase tends to predominate in lentigo-maligna melanoma, superficial spreading melanoma and acral lentiginous melanoma. In these lesions, the malignant melanocytes tend to spread horizontally through basal layer of the epidermis.

- *Vertical growth phase*—the vertical growth phase begins when neoplastic cells populate the underlying dermis. With nodular melanoma, the radial growth phase is very short or nonexistent and the vertical growth predominates.

Types

- *Based on clinical features*—the clinical appearance of the tumor is variable and may be divided into the following five types based on clinical features:
 - Pigmented nodular type
 - Nonpigmented nodular type
 - Pigmented macular type
 - Pigmented mixed type
 - Nonpigmented mixed type.
- *Clinicopathologic types*—four clinicopathologic (skin) types of melanoma have also been described.
 - *Superficial spreading melanoma*—superficial spreading melanoma is the most common form of melanoma. It accounts for 65-70% of cutaneous melanomas. It exists in a radial growth phase which has been called as premalignant melanosis or pagetoid melanoma in situ.
 - *Nodular melanoma*—nodular melanoma represents 13-15% of cutaneous melanomas. It apparently has no clinically recognizable radial growth phase, existing solely in a vertical growth phase.
 - *Lentigo-maligna melanoma*—it accounts for 5-10% of cutaneous melanomas. It exists in a radial growth phase which is known as lentigo-meligna or melanotic freckle of Hutchins. It predominantly occurs on sun exposed skin of elderly person. It is characterized by an extensive radial growth phase which may last up to 25 years before connective tissue invasion takes place.
 - *Acral lentiginous melanoma*—It is the most common form of melanoma in blacks and also the most common form of oral melanoma. It typically develops on the palms of the hands, soles, feet, subungual area and mucous membrane.
- *Superficial spreading melanoma*—it develops over several years into a sharply outlined, slightly elevated pigmented patch. The lesion presents as a tan, brown, black or admixed lesion on sun-exposed skin, especially in blacks. The most common sites of origin are the intercapsular area of males and the back of the legs of females.
- *Nodular melanoma*—clinically, it appears as darkly pigmented elevations on skin or mucous membrane. Nodular melanoma is usually a deeply pigmented exophytic lesion, although sometimes the melanoma cells are so poorly differentiated that they no longer can produce melanin, resulting in a nonpigmented amelanotic melanoma. This form of melanoma is invasive in nature from the start and metastasizes early. It has relatively poor prognosis.
- *Lentigo-maligna melanoma*—clinically, it appears as a flat, irregularly pigmented patch with an ill-defined margin. The appearance of nodularity with a lentigo-maligna signals the onset of the invasive or vertical growth phase.
- *Acral lentiginous melanoma*—it begins as a darkly pigmented, irregularly marginated macule which later develops a nodular invasive growth phase.

Oral Manifestations

- *Site*—oral melanoma exhibits definite predilection for the palate and maxillary gingiva.
- *Color and appearance*—an oral lesion typically begins as a brown to black macule with irregular borders. The macule extends laterally and a lobulated exophytic mass develops once the vertical growth is initiated.
- *Signs*—ulceration may develop early (**Fig. 20-8**) but many lesions are dark, lobulated, exophytic masses without ulceration at the time of diagnosis.



Fig. 20-8: Malignant melanoma showing ulceration and deep color pigmentation in the palate.

Clinical Features

- *Age and sex*—it can occur at any age group, but it is extremely rare in ages below 30. The age range for patient with oral melanoma is from 40-70 years, the average age being 55 years. It is more common in men.
- *Site*—the most frequent oral sites of occurrence are the palate and the maxillary gingiva, accounting for 80% of all melanomas. The most frequent head and neck site of occurrence of mucosal melanoma are the conjunctiva, followed by the upper respiratory tract and the oral cavity.

- *Symptoms*—pain is not a common feature except in ulcerated lesions and most lesions are relatively soft on palpation.

Radiological Features

- *Site*—in most cases, only the soft tissues are affected. Both primary and secondary malignant melanoma may rarely involve the bone of the jaws.
- *Moth eaten appearance*—some melanomas in the jaws, like those in other bones, may present radiographic appearances which are almost indistinguishable from osteomyelitis and patients have even been treated for this condition, until with the passage of time the error becomes apparent. Underlying or adjacent bone may show radiographic evidence of irregular or “moth-eaten” destruction.

Differential Diagnosis

- *Oral melanotic macule*—because of many clinical similarities exist between melanoma and its counterpart, the melanocytic nevus, an “ABCD” system of evaluation has been developed to help distinguish a melanoma clinically from a melanocytic nevus. **A**symmetry (because its uncontrolled growth pattern), **B**orders irregularity (often with notching), **C**olor variation (which varies from shades of brown to black, white, red and blue, depending upon amount and depth of melanin pigmentation) and **D**iameter often greater than 0.5 cm.
- *Melanotic neuroectodermal tumor of infancy*—occurs in infants.
- *Ecchymotic patch*—it disappears within few days.
- *Melanoplakia*—it is firm and occurs in infancy or puberty without change.
- *Intramucosal compound or blue nevus*—same as oral melanotic macule.
- *Pigmented fibroma*—histopathological diagnosis should be done.

Management

- *Surgical excision*—the treatment for cutaneous malignant melanoma is surgical excision. Although regional lymph node dissection is indicated when nodes are involved; prophylactic lymph node dissection is very controversial.
- *Radiotherapy*—radiotherapy is applied mainly on patients unable to undergo surgical treatment or as an adjuvant to surgery.
- *Preoperative chemotherapy*—preoperative chemotherapy is occasionally used to reduce the size of the melanoma and improve surgical management. The first line chemotherapeutic agent is dimethyltriazeno, imidazole carboxamide solely or in combination with vincristine

or dactinomycin. Other immunotherapeutic drugs that are occasionally used include interferon and cimetidine which when used together are believed to activate killer T-cells and inhibit suppressor T-cells resulting in the reduction of the size of the melanoma.

- *Intralesional injection*—Intralesional injection of interferon and polyvalent melanoma antigen vaccine.

Drug-induced Pigmentation

A number of medications may induce oral mucosal pigmentation. Direct deposition on oral surfaces, local accumulation after systemic absorption, stimulation of melanin related pathways, bacterial metabolism, alone or in combination, may result in oral pigmentations. Several antimalarial drugs are known to be capable of inducing intraoral melanin pigmentation. These drugs include: quinacrine, chloroquine, and hydroxychloroquine. Long term use may cause pigmentation of the oral mucosa.

Clinical Features

- *Color of pigmentation*—the pigmentation of the oral mucosa is described as slate-gray in color, bearing some resemblance to pigmentation caused by silver arsphenamine.
- *Location*—they are usually located on hard palate. Amodiaquine has been used to manage autoimmune disease and oral pigmentation occur in 10% of patient taking the drug on long term basis.
- *Minocycline pigmentation*—minocycline is a synthetic tetracycline that is commonly used in the treatment of acne vulgaris. Although tetracycline causes pigmentation of bones and teeth, minocycline alone is also responsible for soft tissue pigmentation. It is usually seen as brown melanin deposits on the hard palate, gingiva, mucous membranes, and the tongue.



Fig. 20-9: Drug induced pigmentation showing brown pigmentation.

- *Birth control pill pigmentation*—birth control pill can be associated with brown pigmentation of facial skin and perioral region.
- *Phenolphthalein*—these are associated with well circumscribed area of hyperpigmentation on skin and oral mucosa.

Differential Diagnosis

The diffuse distribution of pigment in drug induced lesion is similar to that seen in Addison's disease and, in the palate, to early superficial spreading melanoma. The history is, of course, of prime importance and biopsy is recommended.

Treatment

Withdrawal of the medication is recommended, although the pigmentation may persist for as long as one year after cessation of medication.

Physiologic Pigmentation

Oral pigmented lesions are usually caused by increased amount of melanin deposition. Physiologic pigmentation is one of these lesions and is defined as localized, symmetric hyperpigmentation which is commonly seen on attached gingiva.

Etiopathogenesis

- *Genetic*—physiologic pigmentation is probably genetically determined.
- *Mechanical, chemical and physical stimulation*—Dummett suggested, that degree of pigmentation is partially related to mechanical, chemical, and physical stimulation.
- *Activity of melanocytes*—in darker skinned people, oral pigmentation increases, but there is no difference in the number of melanocytes between fair-skinned and dark-skinned individuals. The variation is related to differences in the activity of melanocytes.
- *Age factors*—there is some controversy about the relationship between age and oral pigmentation. Steigmann and Amir et al stated that all kinds of oral pigmentation appear in young children. Prinz, on the other hand, claimed that physiologic pigmentation did not appear in children and was clinically visible only after puberty.

Clinical Features

This type of pigmentation is symmetric and persistent and does not alter normal architecture, such as gingival stippling.

- *Age distribution*—this pigmentation may be seen in patient at any age and is without gender predilection.

- *Location*—it may be found in any location, although the gingiva is the most commonly affected intra oral tissue. The most common pattern of physiologic pigmentation is the band of pigments on attached gingiva. Pigmentation may also be present on the buccal mucosa.
- *Color*—The color of the lesions ranges from brown to black to blue, depending on the amount of melanin production and the depth/location of the pigment.
- *Tongue pigmentation*—the tongue, however, is infrequently pigmented, but when this occurs, the pigments are characteristically localized at the tip of isolated groups of filiform papillae. Pigmentation was rarely found on the ventral surface of the tongue (**Figs 20.10 and 20.11**).
- *Post inflammatory pigmentation*—a related type of pigmentation, called postinflammatory pigmentation, is occasionally seen following mucosal reaction to injury.



Fig. 20-10: Physiologic pigmentation seen on tongue of patient.



Fig. 20-11: Physiologic pigmentation seen on tongue.

Differential Diagnosis

Clinical differential diagnosis would include smoking associated melanosis, the syndrome associated with oral

melanosis, Peutz-Jegher's syndrome and Addison's disease and melanoma. Although physiologic pigmentation is usually clinically diagnostic, biopsy may be justified if clinical features are atypical.

Treatment

- **Gingival surgery**—melanin pigmentation of the oral tissue does not present a medical problem, but some patient may complain of esthetic problem due to black gum. To overcome this issue, gingival surgery might be suggested.
- **Cryotherapy**—recently, cryotherapy is suggested for removal of such type of pigmentation has more advantages, like quickness, simplicity, lack of bleeding and scar compared to other method.

Post-inflammatory Pigmentation

Long-standing inflammatory mucosal diseases, particularly lichen planus (Figs 20-12 and 20-13), can cause mucosal pigmentation.

Etiology

The pathogenesis of post-inflammatory pigmentation remains unclear.



Fig. 20-12: Pigmented lichen planus present on buccal mucosa.

Clinical Features

This is seen more frequently in dark-skinned individuals. Clinically, multiple brown-black pigmented areas are noted adjacent to reticular or erosive lesions of lichen planus.

Cyanosis

- **Cause**—it is caused by substantial decrease in proportion of reduced hemoglobin to oxygenated hemoglobin in blood. It becomes apparent when reduced Hb is less than 5 g/100 ml.



Fig. 20-13: Pigmented lichen planus showing dark pigmentation.

- **Color**—generalized bluish cast to skin is seen in light skinned individuals. Generalized green may be caused by congenital heart defect, right ventricular failure and ventricular fibrillation and is associated with clubbing. Cold weather may cause peripheral cyanosis on lip.

HIV Oral Melanosis

- **Site**—buccal mucosa is most frequently affected followed by gingiva, palate and tongue.
- **Color**—it presents with diffuse, multifocal, macular brown pigmentation of the buccal mucosa.

Addison's Disease

It is also called as '*chronic adrenal insufficiency*'. Addison's disease is a hormonal disorder resulting from a severe or total deficiency of the hormones made in the adrenal cortex. Addison's disease is relatively uncommon condition estimated to occur in 1 in 100000 of the population. The oral manifestation has been documented, but it is unclear in how many cases oral pigmentation without systemic upset or skin pigmentation is the presenting feature. Clinically the disease is characterized by a bronzing of the skin and a pigmentation of the mucous membrane. Both oral and skin pigmentation are thought to be result from melanocytes-stimulating hormonal activity.

Etiology

- **Tuberculosis**—it was the leading cause of Addison's disease until the antibiotics were introduced that successfully treated TB.
- **Autoimmune disorders**—nowadays, the major cause is an autoimmune disorder in which the body's immune system makes antibodies which attack the cells of the adrenal cortex and slowly destroys them. This can take months to years.

- *Adrenocortical destruction*—bilateral adrenocortical destruction after tuberculosis or fungal infection and an idiopathic atrophy are the most frequent causes.
- *Others*—occasionally, bilateral tumor metastasis, leukemic infiltration, and amyloidosis of the adrenal cortex have been found to be responsible. Other less common causes include other chronic infections, cancer that has spread to the adrenals, CMV virus, and surgical removal of the adrenal glands. Whatever the cause, the loss of adrenal cortex results in deficiency in both glucocorticoids and minerlocorticoids.

Pathogenesis

It has been shown that ACTH and melanocytes stimulating hormone (MSH) are similar in structure, and ACTH is believed to have some degree of melanocytes stimulating activity. Normally, the pituitary gland produces ACTH which causes the adrenal cortex to produce glucocorticoids (such as hydrocortisone), which in turn are secreted into the circulation. When the glucocorticoids reach a certain concentration in the blood, they cause the anterior pituitary to cease production of the ACTH. In Addison's disease, however, the defective cortex is unable to produce much glucocorticoid, so this feedback mechanism is not activated and the pituitary continues to produce ACTH. As a result of the increased production of melanin changes, the color of the skin is a smoky tan or a chestnut brown.

Clinical Features

- *Signs and symptoms*—the signs and symptoms of Addison's disease are generally non-specific and include fatigue, weakness, weight loss, nausea, abdominal pain, diarrhea, and vomiting and mood disturbances. These symptoms steadily worsen over time due to the slowly progressive loss of cortisol and aldosterone production.
- *Skin signs*—The following skin signs may be indication of Addison's disease and should take prompt further investigations and appropriate tests.
 - Generalized darkening of the skin (hyperpigmentation) that may look like an inappropriate tan on a very ill person.
 - Hyperpigmentation is most evident on areas exposed to light, but also affects the body folds, sites of pressure and friction, and in the creases of palms and soles
 - Hyperpigmentation may also appear prominent on the nipples, armpits, genitals and gums (buccal mucosa).
 - Women may have loss of androgen-stimulated hair, such as pubic and underarm hair
- *Pigmentation*—pigmentation usually appears early and is one of the most prominent signs of the disease.

It may take one of the two forms, the more usual being a deep tanning of the skin and mucous membrane with heavier deposits of melanin over pressure points. The cheek is the most common site for this pigmentation in the oral mucosa. More infrequently, the increased melanocytic activity is expressed by the development of distinct brownish macule on the oral mucosa and the skin.

- *Color of pigmentation*—bluish black to pale brown or deep chocolate spreading over the buccal mucosa from the angle of the mouth (**Fig. 20-14**) or developing on the gingiva, tongue and lips.
- *Diagnosis*—if Addison's disease is suspected, tests measuring cortisol and aldosterone blood and urine levels must be performed to make a definitive diagnosis. The diagnosis of the Addison's disease is based on the clinical sign as well as on characteristic changes in the blood sodium and chloride levels.



Fig. 20-14: Bluish black pigmentation seen on buccal mucosa in Addison's disease.

Differential Diagnosis

- *Hyperpituitarism*—Addison's disease may be distinguished from hyperpituitarism by the use of urine test: levels of 17-ketosteroids in the urine are decreased in the former but elevated in latter condition. A history of silver ingestion identifies argyria.
- *Peutz-Jegher's syndrome, Albright's syndrome and von Recklinghausen's disease*—the macular type of discoloration that occasionally develops in place of the more generalized tanning might be mistaken for Peutz-Jegher's syndrome, Albright's syndrome, or von Recklinghausen's disease; however, the attending feature these individual syndromes should preclude any such confusion.

Management

It is done by adequate corticosteroid maintenance therapy provided by an average daily dose of 25 to 40 mg cortisone.

Peutz-Jegher's Syndrome

It is also called as *hereditary intestinal polyposis syndrome*. Peutz-Jegher's syndrome (PJS) is a rare familial disease first described by Peutz in 1921 and Jegher 1949 is an autosomal dominant disorder featuring gastrointestinal polyp and the pigmented macule on the lip and skin.

Etiology

To date, it has not been possible to determine the gene, although germline mutations have been detected on the LKB1 (STKI11) gene on chromosome 19p13.3. These are nonsense, frameshift and mis-sense mutations and all inactivate LKB1 gene which encodes a threonine kinase and behaves like a tumor suppressor gene.

Inactivation of this gene by germline mutations or loss of normal allele can result in hamartomatous polyps. Apart from the role of beta-catenin, adenomatous polyposis coli, K-ras and p-53 gene mutations have also been investigated.

The mucocutaneous pigmentations caused by melanin aggregation can be seen in 93% of patients even in infancy. They are generally located around the mouth, nose, cheek mucosa, hand, foot and sometimes perianal and genital areas.

Clinical Features

- **Age**—generally, it is of childhood onset with no sex predilection.
- **Intestinal polyps**—the major manifestation of PJS is the intestinal polyps that are found anywhere in the gastrointestinal tract, but most frequently in the small bowel especially in the jejunum. The number and the rate of growth of these polyps are variable.
- **Symptoms**—the polyp usually becomes symptomatic between the ages of 10 and 30, and may cause ulceration with bleeding, obstruction, diarrhea, and intussusceptions. Patient may be anemic from chronic blood loss. The polyps are usually benign, but malignant transformation may occur. Peutz-Jegher's syndrome has been accepted as a precancerous syndrome, with cancers being seen in 50% of patients.
- **Tuberous sclerosis**—tuberous sclerosis is a frequent neurologic problem in PJS patients and it is characterized by hamartomatous polyp, mental retardation, and epilepsy and adenoma sebaceum.
- **Pigmentation**—it is also characterized by multifocal macular melanin pigmentation in perioral location. It manifests itself as freckle like macules about the hands,

perioral skin (**Fig. 20-15**), and intraorally to include the gingiva, buccal, and labial mucosa. Pigmented spots are 1 to 10 mm in diameter and asymptomatic.

- **Sites of oral pigmentation**—pigmented spots are particularly found on the lower lip and buccal mucosa but rarely on the upper lip, tongue, palate, and gingiva. The pigmented macules appear at birth or in early childhood and may gradually disappear from the skin but not from the buccal mucosa. PJS is easily distinguished from freckles which are usually on sun exposed area and from generalized lentigines which are widely scattered. The pigmentation of the PJS, on the other hand, is acral, perioral, and oral.
- **Color of pigmentation**—there are *bluish-black* macules on skin. Orally it presented as brownish macules.
- **Clinical significance**—the lesion is of clinical significance only as a diagnostic clue to the more serious intestinal problem. Any child with recurrent unexplained abdominal pain should be examined for the cutaneous and mucosal pigmentation of PJC. It is recommended that any patient presenting with ileus attacks and anemia should be investigated for polyps and the mucocutaneous pigmentation of Peutz-Jeghers syndrome. In addition, patients in whom this syndrome is diagnosed should be evaluated for cancer and family screening should be considered.



Fig. 20-15: Macular pigmentation seen in Peutz-Jegher's syndrome.

Management

Referral to a gastroenterologist is recommended. No treatment is required for oral lesions.

Neurofibromatosis

It is also called as *Von Recklinghausen's disease*. It is transmitted as autosomal dominant trait. In it, multiple neurofibromas of the skin along with brown spots are seen.

Clinical Features

- **Triad**—clinical triad consists of areas of pigmentation (**Fig. 20-16**) sessile or pedunculated tumors of skin and mucous membrane and tumors of nerves.
- **Tumors**—tumors are of plexiform variety and thus are soft, smooth, fluctuant, flesh colored and nodular or pedunculated. Multiple cavernous neurofibroma and auxiliary folding can be seen.
- **Café-au-lait spots**—are cutaneous lesions which are characteristic of the disease.



Fig. 20-16: Area of pigmentation seen with swelling on the face.

Oral Manifestation

- **Site**—areas of melanin pigmentation are seen on oral mucosa with lips being the common site of involvement.
- **Intraoral growth**—there is presence of intraoral growth inside the oral cavity (**Fig. 20-17**).



Fig. 20-17: Intraoral sessile growth seen in patient of neurofibromatosis.

Radiographic Features

- **Appearance of intrinsic bone tumor**—in intrinsic bone tumors, there are actual tumor masses within the bone which appear as central cyst like radiolucency, subperiosteal bone blister and irregular areas of bone destruction.
- **Appearance of extrinsic bone tumor**—extrinsic bone tumor produces an area of bone destruction or prevents normal development of contour of the bone. Tumor which affects only the superficial portion of the bone may result in the formation of depression and elevation, pits and superficial cavities.

Differential Diagnosis

- **Basal cell syndrome**—tumor is restricted to skin of face, neck and chest. Pigmentation is not feature.
- **Gardener's syndrome**—pigmentation is not seen.

Management

- **Surgical excision**—surgical excision of individual lesion.

Albright's Syndrome

- A type of fibrous dysplasia involving all the bones in the skeleton accompanied by the lesions of the skin and endocrine disturbances of varying types.
- Skin lesions are irregularly pigmented melanotic spots described as café-au-lait spots due to their light brown color.
- It is more common on lips.

Differential Diagnosis

- **Addison's disease**—discrete macules are seen in adrenal insufficiency.
- **Peutz-Jegher's syndrome**—pigmented macules on skin restricted to areas surrounding the body orifices, on finger and both.

Hyperfunction of the Pituitary Gland

There is increased secretion of ACTH and MSH thus causing increase in melanization rate causing pigmentation.

Pregnancy and Female Sex Hormones

Increased ACTH levels during pregnancy is the cause of abnormal pigmentation of circumoral tissues and nipples are seen in the third trimester of pregnancy, this is called as *chloasma grandarum*. This also occurs with use of oral contraceptive containing large doses of sex hormones. In all these, functions of pituitary gland are increased causing pigmentation.

Occasionally, it is accompanied by diffuse browning of oral mucosa and slowly disappears after delivery.

Brown Heme Associated Lesion

Ecchymoses and Petechiae

They are purpuric submucosal and subcutaneous hemorrhages (Fig. 20-18). Petechiae are minute pinpoint hemorrhages while ecchymosis is larger than 2 cm in diameter.



Fig. 20-18: Petechiae seen on tongue due to trauma.

Clinical Features

- **Site**—common on lips and face.
- **Causes**—it occurs immediately following traumatic event and erythrocyte extravasation into submucosa.
- **Color**—it appears as bright red macule (Fig. 20-19) or swelling, if hematoma is formed. After hemoglobin is degraded to hemosiderin, lesion assumes brown color. It does not blanch on pressure.
- Fluctuant swelling, as hemorrhage is slow and no sufficient blood to pool.

Differential Diagnosis

- *Amalgam tattoo, oral melanotic macule, junctional nevus, melanoma and telangiectasia in Rendu-Osler-Weber syndrome*—in all of the above, history of recent trauma, change from bluish brown to green to yellow and then disappearing lesion within 4 to 5 days indicate petechiae.
- *Trauma from fellatio*—disappear in few days after passing through blue to green to yellow color changes to be taken. Careful history should be taken.
- *Trauma from vomiting and coughing*—broad linear red or bluish bruise.



Fig. 20-19: Leg macule seen as bright red macule.

- *Infectious mononucleosis*—malaise, enlarged nodes in neck, positive monospot test followed by positive Paul Bunnell heterophil test.
- *Hemostatic disease*—complete blood examination is done.

Management

- Surgery, after detection of disease.

Hemochromatosis or Bronze Diabetes

- **Tetrad**—it is tetrad of liver cirrhosis, diabetes, cardiac failure and bronze skin.
- **Causes**—Bronze diabetes (Hemochromatosis) is a disorder in which excess iron is deposited in the body and result in eventual sclerosis and dysfunction of the tissue and organ involved. The cause of tanning in hemochromatosis, like that in Addison's disease, is an increased melanin production and not the deposition of hemosiderin in the skin. This increased production, as in Addison's disease result from the high level of ACTH that accompany the destruction of the adrenal cortex by the heavy iron deposits.
- **Site**—Involved organs are liver, skin, pancreas and adrenal gland.
- **Sex distribution**—80% occurs in men
- **Color**—tanning occurs due to increased melanin production. Blue gray color of skin especially over genitals, face and arms.
- **Intraoral features**—in this, diffuse black brown pigmentation is seen at the junction of hard and soft palate (Fig. 20-20).

Carotenemia

Causes—chronic excessive levels of carotene pigments due to long and continuous consumption of food containing



Fig. 20-20: Black brown pigmentation seen at the junction of hard and soft palate in hemochromatosis.

carotene such as carrots, sweet potatoes and egg. An orange to yellow pigmentation of skin and oral mucosa occurs. Color change is more intense on palm, soles and areas of soft palate.

Jaundice

Yellow or green discoloration of the skin and mucous membrane with bile pigments. Oral mucosa presents a yellowish discoloration. Tongue is heavily coated.

Hematoma

Hematoma is a localized collection of blood, usually clotted, in a tissue or organ. Lingual hematoma has been documented as a result of maxillofacial trauma causing tongue laceration, post-extraction complication, severe hypertension, unusual complications of anticoagulant therapy, and surgical implant placement. Although not recorded in the dental literature, periodontal flap procedures may also affect hematoma formation. While a relatively uncommon sequelae of surgical therapy, the broad spectrum of events resulting in hematoma necessitates the dental practitioner's familiarity with its diagnosis and treatment. The floor of the mouth hematoma can be more problematic because of the facial planes are involved and the anatomical structures at this site.

Etiology

A hematoma is generally the result of hemorrhage, or, more specifically, internal bleeding. Hematoma exists as bruises (ecchymoses), but can also develop in organs. Injuries to the facial bones and jaw can produce bleeding not only in the tongue, but also in the adjacent facial structures. Excessively traumatized capillary beds extravasate blood

into the perivascular connective tissue. Poor circulation at the site allows pooling of blood, prostaglandin release, and a more profound inflammatory reaction.

Clinical Features

Mucosal hematoma bruise result from vascular severance result from trauma (**Fig. 20-21**). They are brown or blue and may be macular or swollen. Because the blood is intraluminal but extravasated, hematoma does not blanch on pressure. The early hematoma is fluctuant, rubbery, and discrete in outline, and the overlying mucosa is readily movable. The temperature of the overlying mucosa may be slightly elevated. When it is superficial, it appears as an elevated bluish swelling in the mucosa. Digital pressure on the surface may induce a stinging sensation as the pressure on the contained pool of the blood causes further separation of the tissue. A hematoma is usually completely clotted within 24 hours and then becomes a hard black, painless mass. If the hematoma is superficially located, changes from black to blue to green to yellow may be observed during the following days. If hematoma becomes infected, it is painful. Although the clot is initially firm, if the infection is a pyogenic type, the firm clot softens and become fluctuant as pus accumulates.

Hematoma involving the tongue frequently affect swallowing and speech. As the hematoma enlarges, or extends into the surrounding tissue below the tongue, obstruction of the airway occurs.



Fig. 20-21: Hematoma seen on tongue due to trauma.

Differential Diagnosis

A history of traumatic incident like accident, surgery, or administration of local anesthetic can almost always be elicited from the patient and is useful in establishing the diagnosis of early hematoma. Hematomas are the result of trauma may be confused clinically with hemorrhagic mucocoele, tattoo, hemangioma or varix, nevus, melanoma.

Ecchymosis is identical, yet in blood dyscrasia, trauma is minor and the lesions are usually multifocal

Treatment

Hematomas that occur intracranial require immediate specialized medical attention. The hematoma is usually self-limiting in size because the increasing pressure of the blood in the tissue equalizes with the hydrostatic pressure in the injured vessel and thus terminates the extravasations. If a large arteriole is damaged, however, a pressure bandage may be placed over the accessible area to control the hemorrhage and limit the expansion of the hematoma. Occasionally, it may be feasible to evacuate an expanding or painful hematoma with an aspirating syringe and then apply a pressure bandage to prevent its re-formation. If the patient appears to be developing a problem with respiration the patient should be hospitalized and proper measure instituted to establish and maintain the airway. Antibiotic may be given to prevent the secondary infection.

For contusions (bruises), treatment consists of initially applying ice or cold packs a few times a day, to produce vasoconstriction (a reduction in arterial blood flow) which helps to decrease hemorrhage (bleeding) and edema (swelling). A suspected hematoma should be observed for 2-week period. Failure to resolve indicate the clinical impression was incorrect and biopsy should be performed. Any hematoma observed without significant provocation or trauma should arouse suspicion of purpura associated with blood dyscrasia.

Exogenous Pigmentation

It is defined as a pigmentation which arises as a result of introduction of metal/drugs into the body via mucous membrane, intestinal tract and skin.

Classification

- *Accidental pigmentation*—this type of pigmentation occurs due to accidental implantation of material in the gingival tissue.
- *Iatrogenic pigmentation*—this occurs due to dental procedure.
- *Heavy metal pigmentation*—this occurs due to ingestion of metal in the body.

Accidental Pigmentation

Foreign substances are embedded in the gingival tissue. It may be due to:

- *Accident during childhood*—for example, if one falls on road, some particles of the road surface gets embedded in gingiva and if not removed, may cause discoloration.
- *Charcoal tooth powder*—charcoal containing tooth powder also produces black permanent discoloration.
- *Graphite tattoos*—pencil points are occasionally broken off in gingival tissue and if not completely removed, may cause permanent discoloration. It tends to occur on the palate. The lesions are usually macular, focal, and gray or black. Since the traumatic event usually occurs in classrooms during grade school, many patients may not recall the injury. Pigment does not blanch on pressure. History will confirm the diagnosis of graphite tattoo.

Iatrogenic Pigmentation

Amalgam Tattoo

It is also called as *localized argyrosis*. Dental amalgam tattoos are relatively common intraoral pigmented lesions, with an estimated incidence of 8%. It is characterized by the deposit of restorative debris composed of a mixture of silver (Ag), mercury (Hg), tin (Sn), zinc (Zn), and copper (Cu) in subepithelial connective tissue.

Etiology

- *Restorative work*—it may be condensed in the abraded gingiva during routine amalgam restorative work.
- *Removal of old filling*—it may enter the mucosa lacerated by rotary instruments during removal of old amalgam fillings or crown and bridge preparations of teeth with large amalgam restorations.
- *During extraction*—broken pieces may be introduced into the socket or beneath the periosteum during extraction of the teeth.
- *Retrograde amalgam filling*—particles may enter the surgical cut during root canal treatment with retrograde amalgam filling. There may be chances of corrosion of retrograde amalgam filling.

Clinical Features

- *Site*—the most common sites are on gingiva and alveolar mucosa with mandibular region being affected more commonly than maxillary region.
- *Age and sex distribution*—it can occur at any age but it is rarely seen below the 12 years as amalgam restorations are not used before the age of 12 years. Females are affected more commonly than males in ratio of 1.8:1.
- *Appearance*—it is described as a flat macule or sometimes slightly raised lesion with margins being well defined or diffuse in other.
- *Color*—pigmentation is blue black in color (Fig. 20-22). It may gradually increase in size.



Fig. 20-22: Blue black pigmentation seen adjacent to amalgam restoration in amalgam tattoo (Courtesy Dr Chole).

Radiological Features

- It will demonstrate the presence of the metal.

Differential Diagnosis

- *Superficial hemangioma*—it blanches on pressure while amalgam tattoo does not.
- *Nevus and melanoma*—rare in oral cavity. It has brown color as compared to tattoo which has blue black color.

Management

- *Surgical excision*—once present, the amalgam tattoo remains indefinitely, and occasional lesions slowly enlarge over time, presumably as amalgam-laden histiocytes try to move the material out of the local site. No treatment is necessary, but excisional biopsy is often performed in order to rule out melanoma or another pigmented lesion. Lesions visible on radiographs are usually not biopsied and those occurring on the visible vermillion border of the lips are usually removed for aesthetic reasons. There is no malignant potential for this lesion.

Pigmentation due to Drugs and Metals

Bismuthism

It is systemic metallic intoxication due to bismuth compound.

Etiopathogenesis

- *Medicinal use*—nowadays, medicinal use of bismuth is rare. In the past, it is used in the treatment of venereal disease. Many proprietary drugs contain bismuth salt and bismuth containing pastes may result in bismuth pigmentation.
- *Occupational exposure*—person working in bismuth factory can cause bismuth intoxication.
- *Mechanism*—pigmentation is produced by the action of hydrogen sulfide on the bismuth compound. The hydrogen sulfide is formed through bacterial degradation of organic material of food retention.

Clinical Features

- *Symptoms*—vague gastrointestinal tract disturbances, nausea, bloody diarrhea, *bismuth gripe* and jaundice.
- *Bismuth line*—sometimes in the long bone, white bands of increased density appear in the ends of the diaphyses immediately adjacent to the epiphyseal lines. This is called as 'bismuth line'.

Oral Manifestations

- *Symptoms*—patients often complain of a metallic taste, increase salivation with burning sensation in the oral cavity
- *Ulcerative gingivostomatitis*—it is very common occurrence in bismuth poisoning.
- *Sign*—large, extremely painful, shallow ulcerations are seen at times on the cheek mucosa in molar region. Regional lymphadenopathy may be present.
- *Tongue*—tongue is frequently enlarged and sore.
- *Bismuth line*—'blue black' bismuth line appears to be well demarcated to eye on gingival papillae. Blue black bismuth sulfide granules formed by action of H_2S produced by action of bacteria on organic material remaining in areas of poor oral hygiene.
- *Paper test*—it will indicate whether the pigmentation is actually in gingival tissue. If the pigmentation persists, when small piece of white paper is inserted in the gingival sulcus, the presence of pigmented area is verified.

Management

- *Removal of cause*—Stoppage of use of bismuth.
- *Oral hygiene maintenance*—dentist should establish and maintain oral hygiene of the patient.
- *Topical anesthetics*—management of painful ulcerative lesions should be done by topical application of lignocaine hydrochloride gel.

Plumbism or Lead Poisoning

It occurs due to lead poisoning.

Etiopathogenesis

- **Lead based paint**—lead is present in paint and children can ingest chips of paint. While renovation and sanding, lead may appear in dust resulting in lead intoxication.
- **Lead in illicit alcohol**—moonshine an illicit alcoholic beverage distilled in car radiators has been shown to cause acute lead poisoning.
- **Lead in gasoline**—use of tetraethyl lead, an antiknock compound in gasoline (petrol), has introduced a new source of lead to the public. Removal of lead from gasoline is nowadays carried out and you can get unleaded petrol.
- **Occupational exposure**—excessive absorption of the lead from automobile exhaust and dust can lead to occupational exposure of lead.
- **Mechanism**—absorption of lead from alimentary tract, lungs and gut → modulated by vitamin D and calcium status of the individual → lead is taken up by circulating erythrocytes and bound to reactive sulfhydryl group of proteins → from the circulation, lead is transferred to all the soft tissues and in high concentration, it will inhibit metabolic pathways → in the red cells, lead inhibits enzymes associated with hemoglobin synthesis, hence abnormal activity of the enzymes occurs.

Clinical Features

- **Nervous system**—lead has high affinity for cells in central as well as peripheral systems. In acute poisoning, demyelination and axon degeneration occurs. Lead encephalopathy, cerebral palsy, mental retardation, seizures, wrist or foot drop and fatigue can occur.
- **Gastrointestinal tract**—there may be serious gastrointestinal disturbances like nausea, constipation, vomiting, and colic.
- **Bone**—when incorporated in the bone, it can interfere with cellular metabolism and changes are seen in the rate of bone resorption and apposition.

Oral Manifestations

- **Symptoms**—there is a metallic taste which is accompanied by excessive salivation and dysphagia.
- **Burtonian line**—when exposure to lead is very high and oral hygiene is very poor, a line known as 'burtonian line' is seen which is *gray black* in color (**Fig. 20-23**) and is present along the gingival margin. Lead line is more diffuse than bismuth line.
- **Signs**—there is pallor of lip, poor muscle tone and the face appear *ashen* in color because of associated anemia. Tremor of tongue may present on thrusting.
- **Parotid gland**—there is bilateral parotid gland hypertrophy.



Fig. 20-23: Plumbism showing gray black line on the gingiva.

Radiological Features

- **Lead line**—lead line can be seen in the long bones and skull. It represents deranged calcium metabolism that results from the activity of lead on osteoclastic activity and calcium resorption.

Management

- **Chelating agents**—lead can be removed from body by using a chelating agent such EDTA (calcium disodium ethylenediaminetetraacetate). But nowadays, other chelating agents DMSA (2, 3-dimercapto-succinic acid) and DMPS (2, 3-dimercaptopropane-1-sulfonate) are used.

Mercurialism

It is also called '*Pink disease*', '*Swift's disease*', '*Dermatopolyneuritis*', and '*Acrodynia*'. It is an uncommon disease caused due to a mercurial toxicity reaction, either actual mercury poisoning or, more likely, an idiosyncrasy to the metal.

Etiology

- **Medicinal use**—mercury can be used in teething powder, cathartic agents and antihelminthics preparation.
- **Mercury in dental amalgam**—improper use of dental amalgam alloy which contain mercury can also cause mercurialism. But there is no documented literature on the mercury toxicity occurring due to dental amalgam.
- **Mercury in paint**—mercury hazards exist in paints containing mercurial salts such as phenylmercuric propionate.
- **Mercurial diuretics**—prolonged administration of mercurial diuretics can also result in mercurialism.

- *Night cream*—frequent use of night cream containing inorganic salts may produce distinctive discoloration.
- *Occupational*—mercurial fumes which are produced industrially also result in bone changes in the jaws, if inhaled in quantity.

Clinical Features

- *Age*—it occurs most frequently in young infants before the age of 2 years although children can be occasionally affected up to age of five years or six years.
- *Gastrointestinal symptoms*—intestinal colic and diarrhea. There is also pharyngitis, dysphagia, nausea, abdominal pain.
- *Nervous symptoms*—long continued exposure to mercury vapor can result in permanent neurological changes. Headache, insomnia, tremors of fingers and tongue and mental depression.
- *Renal symptoms*—severe intoxication and it can be the cause of death.
- *Color of pigmentation*—hands, feet, nose and cheeks assume pink color.
- *Hair and nails*—the nails are shed at the same time with teeth lost prematurely and alopecia is also present. The children will frequently tear their hair out in patches.
- *Raw beef appearance*—the skin of hands, feet, nose, ears and cheek becomes clammy red or pink and has a cold clammy feeling. The appearance is described as resembling raw beef.
- *Skin*—the skin over the affected area peels frequently during the course of the disease. The patients also have maculopapular rash which is extremely pruritic.
- *Others finding*—severe sweating, extreme irritability, photophobia with lacrimation, insomnia, muscular weakness, tachycardia and hypertension.

Oral Manifestations

- *Symptoms*—there is marked increase in inflow of ropy viscid saliva. This will cause dribbling. Patient may experience itching sensation and metallic taste in the oral cavity. Mastication is difficult due to pain.
- *Gingiva*—the gingiva becomes extremely sensitive or painful and it may exhibit ulceration. The gingiva may become blue gray to black in color
- *Ulcerative stomatitis*—oral mucosal ulceration occurs and spreads to the palate, throat and pharynx.
- *Salivary gland and lymph nodes*—salivary glands and lymph nodes may be swollen.
- *Tongue*—tongue is enlarged, painful and ulcerated. Tongue tremors may be present.
- *Lips*—lips are dry, cracked and swollen.
- *Teeth*—sound teeth may exfoliate and one or more teeth may be found on bed in the morning as patients awake.

The reason for it is that there is marked periostitis with loosening of the teeth which may lead to exfoliation of teeth. In many cases children extract his own teeth with the help of his finger.

- *Bone*—the loss of teeth sometimes followed by necrosis of bone and there may be sequestrum formation.
- *Bruxism*—bruxism is a common finding.

Radiographic Features

- *Appearance*—changes occurring in the jaw are similar to the changes seen in osteomyelitis except that changes may be localized at first and gradually extend until large portion of bone involved.
- *Irregular bone destruction*—there is irregular area of bone destruction with loss of cortex of the follicle of the affected tooth.

Management

- *Supportive measure*—bed rest and suitable dietary regimen should be adjusted for renal damage.
- *Control of salivary flow*—atropine or belladonna can be prescribed to lessen the salivary flow.
- *Chelating agents*—administration of chelating agents BAL- British anti-lewisite (2,3-dimercaptopropanol) is recommended. But nowadays, as side effect of BAL is more, another chelating agents DMSA (2,3-dimercaptosuccinic acid) and DMPS (2,3-dimercaptopropane-1-sulfonate).

Argyria or Silver Poisoning

It is also called as 'argyrosis' which occurs due to chronic exposure to silver compound.

Etiology

- *Medicinal use*—it occurs from local and systemic absorption of silver compounds. Silver is used in nasal drops or sprays. Silver-arsphenamine injection used to treat syphilis. Silver is also used in some cases of apthous ulcer.
- *Photographic films*—chewing pieces of photographic films over an extended period can also result in argyria.
- *Occupational exposure*—industrial exposure is also one of main cause of silver poisoning.
- *Mechanism*—silver is disseminated in the body and it accumulates as subepithelial deposits in the skin.

Clinical Features

- *Site*—exposed body surfaces, nail-beds are commonly involved.

- **Skin**—skin is slate gray, violet or cyanotic and in marked cases, there is even suggestion of metallic luster.
- **Nails**—nails are also deeply pigmented.

Oral Manifestations

- **Site**—pigmentation is distributed diffusely throughout the gingival and mucosal tissue.
- **Color of gingiva**—there is *slate blue silver* line along the gingival margins.
- **Oral mucosa**—oral mucosa exhibits a diffuse bluish black discoloration.

Management

- **Removal of cause**—source of contact should be eliminated.

Arsenism

It occurs due to arsenic poisoning.

Etiology

- **Industrial exposure**—arsenic exposure from industrial use can lead to arsenic poisoning.
- **Medicinal use**—arsenic is used to treat asthma and dermatose such as psoriasis.

Clinical Features

- **Symptoms**—chronic gastritis and colitis, keratosis of palms of the hand and soles of feet.
- **Hyperpigmentation**—diffuse macular hyperpigmentation is seen on the skin of the patient. This occurs due to increase in melanin production.
- **Arsenical keratosis**—these are the premalignant skin lesion which can occur in arsenic poisoning.
- **Other complication**—patient can also notice palmar and plantar hyperkeratosis, basal cell carcinoma and cutaneous squamous cell carcinoma.

Oral Manifestations

- **Symptoms**—oral tissues are extremely painful and patient may complain of excessive salivation.
- **Severe gingivitis**—gingiva become intensely inflamed.
- **Necrotizing ulcerative stomatitis**—local contact with arsenic trioxide often produces ulceration which can become necrotic.
- **Dorsal hyperkeratosis of tongue**—this occurs in patient in past, who has taken arsenic for syphilis treatment.
- **Color of pigmentation**—tissues are deep red in color.

Management

- **Anesthetic ointment**—surface anesthetic ointment or rinses such as lidocaine or dyclonine solution can be given to control pain occurred due to salivation.

- **Chelating agents**—BAL (2,3-dimercaptopropanol) is used initially for the treatment of arsenic poisoning. But nowadays as side effect of BAL is more another chelating agents DMSA (2,3-dimercapto-succinic acid) and DMPS (2,3-dimercaptopropane-1-sulfonate).

Auric Stomatitis or Gold Poisoning

Auric stomatitis occurs due to gold poisoning.

Etiology

- **Medicinal use**—gold is useful for the treatment of rheumatoid arthritis, lupus erythematosus and leprosy.

Clinical Features

- **Dermatitis**—it is the most common complaint of patient. Patient may notice pruritus before the lesion of dermatitis occurs.
- **Other features**—purpura, alopecia, loss of nails and malignant neutropenia can also occur.
- **Chrysiasis**—slate blue discoloration of skin occur in gold poisoning. This is called as chrysiasis.

Oral Manifestation

- **Site**—it includes buccal mucosa, lateral border of tongue, palate and pharynx.
- **Symptoms**—patient gets metallic test in the oral cavity before the development of stomatitis.
- **Stomatitis**—it is the most common complaint of the patient who is receiving gold therapy.
- **Sign**—vesiculation and ulcerations of the oral mucosa.

Management

- **Removal of cause**—patient should be advised to discontinue of gold therapy.
- **Alkaline mouth wash**—this should be prescribed to treat oral stomatitis.

Pigmentation due to Copper, Chromium, Zinc

- **Copper**—it results due to chronic intake of copper salts which may be associated with development of anemia. There is bluish green line on gingiva and teeth, which is called as 'Clapton line'. Tooth discoloration is permanent because of etching of enamel.
- **Chromium**—chrome platers are exposed to fine spray of chromic acid that is irritating and corrosive to the mucous membrane of the nose and throat. Patient experience burning, soreness, and dryness of mouth associated with swelling of tongue. There is painful ulcerations of the nasal septum which results in perforation. Teeth may become etched and show persistence *deep orange color*.

- **Zinc**—it is an occupational hazard in molten brass workers and electric arc worker. It is associated with chills, fever, sweating and rapid pulse. Nausea, vomiting, dryness, burning of the upper respiratory tract and metallic taste are also associated. In oral cavity, there is congestion and suppuration of the gingival tissues. There are also painful submaxillary lymph nodes and salivary gland involvement also occur. There is *bluish gray line* present on the gingiva.

Summary

Pigmentation is the both normal and abnormal discoloration of oral mucous membrane. Pigmentation has multifactorial etiology. Most of the pigmentation is physiologic but sometimes it can be a precursor of severe diseases. Melanin pigment irregularities and color changes of the oral tissues could provide significant diagnostic evidence of both local and systemic disease. Evaluation of a patient presenting with a pigmented lesion should include a full medical and dental history, extraoral and intraoral examinations, and laboratory tests. The history should include the onset and duration of the lesion, the presence of associated skin hyperpigmentation, the presence of systemic signs and symptoms (e.g. malaise, fatigue, weight loss), use of prescription and nonprescription medications, and smoking habits. Pigmented lesions on the face, perioral skin and lips should be noted. The number, distribution, size, shape and color of intraoral pigmented lesions should be assessed. In general, benign pigmented lesions show regular borders and are small, symmetric and uniform in color. They may be either flat or slightly elevated. In contrast, irregular borders, color variation, and surface ulceration suggest malignancy. Clinical tests such as diascopy, radiography and laboratory investigations such as blood tests can be used to confirm a clinical impression and reach a definitive diagnosis. However, because it is not always possible to distinguish between a benign pigmented lesion and an early melanoma on the basis of clinical features alone, biopsy is usually recommended for focal oral pigmented lesions that cannot be explained by local factors. The recognition, identification, and clinical assessment of pigmentation is of great importance because of the possible risk of serious systemic disease, such as melanoma, various syndromes, and the side effects of drugs.

Hypopigmentation in Oral Cavity

Albinism

The word “albinism” refers to a group of inherited conditions. People with albinism have little or no pigment

in their eyes, skin, or hair. They have inherited altered genes that do not make the usual amounts of a pigment called melanin.

Albinism affects people from all races. Most children with albinism are born to parents who have normal hair and eye color for their ethnic backgrounds. Sometimes people do not recognize that they have albinism.

People with albinism are at risk of isolation because the condition is often misunderstood. Social stigmatization can occur, especially within communities of color, where the race or paternity of a person with albinism may be questioned.

Clinical Features

- **Vision problems**—albinism is associated with vision problems and has low vision. Vision problems in albinism result from abnormal development of the retina and abnormal patterns of nerve connections between the eye and the brain. It is the presence of these eye problems that define the diagnosis of albinism. Therefore, the main test for albinism is simply an eye examination.
- **Skin problems**—while most people with albinism are fair in complexion (**Fig. 20-24**). Skin or hair color is not diagnostic of albinism. People with many types of albinism need to take precautions to avoid damage to the skin caused by the sun such as wearing sunscreen lotions, hats and sun-protective clothing. In less pigmented types of albinism, hair and skin are cream-colored.



Fig. 20-24: Hand of the patient with albinism showing depigmented area.

- **Facial skin**—facial skin show depigmented area (**Fig. 20-25**).
- **Skin cancer**—those who do not use skin protection may develop life-threatening skin cancers.



Fig. 20-25: Hypopigmented region of face in albinism.

Management

- **Skin protection**—use appropriate skin protection, such as sunscreen lotions, higher and opaque clothing, and people with albinism can enjoy outdoor activities even in summer.
- **Avoid social stigma**—families and schools must make an effort not to exclude children with albinism from group activities.

Vitiligo

It is localized type of hypopigmentation. It is progressive symmetric areas of complete pigment loss. It is most commonly seen area around mouth (Fig. 20-26). lip, nose, nipples and anus. Color of pigmentation is chalk white. Management is done by topical glucocorticoids, PUVA therapy.



Fig. 20-26: Hypopigmentation in oral cavity may occur localize type of vitiligo.

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Introduction

Dental caries is a microbial disease of the calcified tissues of the teeth, characterized by demineralization of the inorganic portion and destruction of the organic substance of the tooth. It is one of the most common infectious diseases affecting the human race. Cariogenic plaque contains 2×10^8 bacteria per milligram weight and pH of 5.5 is critical threshold for the demineralization. The initial lesion appears as opaque white or brown spot beneath the plaque layer. As the caries process results in demineralization, the affected area of the tooth appears more radiolucent than unaffected area. Carious area attenuates less radiation than intact tooth substance so that the area of the film on which remnant beam from the demineralized area falls, it receives higher exposure and thus appears more darker on the processed radiograph.

The disease process begins with the concentration of *Streptococcus mutans* at specified tooth surfaces and lead to white spot formation or even cavitations. The development of dental caries is a dynamic process of demineralization of the dental hard tissues by the products of bacterial metabolism, alternating with periods of remineralization.

Etiology

- *Dietary factor*—carbohydrates with types like monosaccharides, disaccharides or polysaccharides and the amount consumed and whether it is between meals.
- *Microorganisms*—acidogenic *Streptococcus mutans* and *Actinomyces viscosus*.
- *Systemic factors*—hereditary, pregnancy and lactation factors have been suggested as etiological factors for dental caries.
- *Host factor*—poor oral hygiene and improper brushing technique can lead to dental caries.

- *Immunological factor*—the functional role of circulating antibodies as protective agents against tooth decay has been demonstrated in non-human primates.

Pathogenesis

- *Fermentation of oral microorganism*—whenever carbohydrate is consumed, oral microorganisms rapidly begin fermentation producing organic acids like lactic acid, acetic acid and formic acid. This leads to fall in pH of the oral fluids.
- *Demineralization*—these organic acid attack the tooth structure, resulting in loss of tooth minerals specially calcium and phosphate ions, which leach out from hydroxyapatite. This process is known as 'demineralization'.
- *Remineralization*—after a period of 30 minutes, due to salivary buffering by bicarbonate ions and ammonia production from salivary proteins, there is an increase in pH of the oral fluids. The acid is neutralized and the condition now favors precipitation of calcium and phosphate ions into tooth surface. This process is called as 'remineralization' and is hastened if fluoride is present in a small amount in either plaque fluid or saliva.
- *Further demineralization*—the microorganism which is of primary concern in the pathology of dental caries is *Streptococcus mutans*. It forms insoluble, sticky extracellular polysaccharides which help in further colonization and increases the contact of the acids with the tooth structure, leading to further demineralization which ultimately leads to cavitations.
- *Formation of caries*—the balance between the caries causing and caries protective factors is very delicate. It is only when repeated attacks of demineralization occur that there is a net loss of minerals from tooth and caries results. The surface layer of enamel overlying the lesion

remains intact and the demineralization occurring is primarily subsurface in location. Once this happens, the process gradually extends deeper, involving enamel and subsequently the dentin and pulp.

Theories of Cariogenesis

Proteolytic Theory

Proteolysis can play a role in dental caries process, particularly in lesions that develop on exposed root surface.

Proteolysis Chelation Theory

It is postulated that oral bacteria attack organic component of enamel and that their breakdown products have chelating ability and this dissolves the tooth minerals.

Chelation is the process involving the complexing of a metallic ion to a complex substance through a coordinate covalent bond which results in a highly stable, poorly dissociated or weakly ionized compound.

Acidogenic Theory

It is generally agreed that dental caries is caused by acid resulting from action of microorganisms on carbohydrates. It is characterized by decalcification of the inorganic portion and is accompanied or followed by a disintegration of the organic substance of the tooth. The cariogenicity of carbohydrate varies with the frequency of ingestion, physical form and chemical composition, route of administration and presence of other food constituents. Sticky solid carbohydrates are more caries producing than those consumed as liquids.

Most commonly associated microorganisms are *L. acidophilus* and *Streptococcus mutans* which are found in caries susceptible individuals. Acids are produced due to enzymatic breakdown of the sugar and the acids formed are chiefly lactic acid and butyric acid.

Autoimmunity

Jackson and Bunch suggest that zones or regions of odontoblasts in specific sites with the pulp of specific teeth are damaged by an autoimmune process so that the defense capacity of the overlying dentin and enamel is compromised and concluded that caries should be regarded as a degenerative process.

Initially disease event corresponds to a form of somatic gene mutation in central growth control stem cells. Descendent mutant cells synthesize autoantibody which damage specific groups of odontoblasts and thus determine the sites of caries susceptibility.

Secondary Factors in Dental Caries

- *Anatomic characteristics of the teeth*
 - *First 2 years after eruption*—teeth are usually susceptible to caries during first 2 years after eruption as additional 2 years are required for completion of calcification after eruption.
 - *First permanent molars*—first permanent molars often have incompletely coalesced pits and fissures that allow the dental plaque material to be retained at the base of the defect in contact with exposed dentin.
 - *Pits on tooth*—lingual pits on the maxillary first permanent molar, buccal pits on the mandibular first permanent molars and lingual pits on maxillary incisors are vulnerable in the process of dental caries proceeds rapidly.
 - *Enamel hypoplasia*—enamel hypoplasia predisposes more to dental caries.
- *Arrangement of the teeth in the arch*—crowded and irregular teeth are not readily cleaned during the natural masticatory process.
- *Presence of dental appliance*—partial dentures, space maintainers and orthodontic appliances often encourage the retention of food debris and plaque material and have been shown to result in an increase in the bacterial population.
- *Saliva factors*—viscosity of saliva has effect on dental caries. Both thick ropy saliva and thin watery saliva have been responsible for rampant caries. Person who has xerostomia is also susceptible for caries.

Classification

First Classification

Based on Location of the Lesion (see Fig. 21-4)

- *Pit and fissure caries*
 - Occlusal
 - Buccal or lingual pit
- *Smooth surface caries*
 - Proximal
 - Buccal or lingual surface
- *Root caries*

Based on Tissue Involved

- Enamel caries
- Dentinal caries
- Cemental caries

Based on Virginity of the Lesion

- Primary caries
- Secondary caries

Based on Progression of Lesion

- Progressive caries
 - Rapidly progressive- like nursing caries and radiation caries
 - Slowly progressive
- Arrested caries

Second Classification

Mount G. J. in 1997 classified dental caries based on site and size.

Site

- *Site 1*—includes lesions on the pit and fissure of the posterior teeth and on other surfaces, these include the buccal grooves of the mandibular molars, palatal grooves of the maxillary molars and erosion on the incisal edges.
- *Site 2*—includes lesions in the contact areas of posterior and anterior teeth.
- *Site 3*—includes lesions originating in the gingival third of all teeth.

Size

- *Size 1 (mild)*—includes lesions which have progressed just beyond remineralization.
- *Size 2 (moderate)*—includes larger lesions with adequate tooth surface to support the restoration.
- *Size 3 (enlarged)*—includes lesions in which the tooth structure and the restoration are susceptible to fracture.
- *Size 4 (severe)*—it includes lesions which have destroyed a major portion of the tooth structure.

Diagnosis of Dental Caries

The search for an ideal caries diagnostic test continues as such test must be accurate, sensitive, specific, reproducible and reliable and should not transfer *S. mutans* or other bacteria from affected area to unaffected areas.

Clinical Method

Visual Inspection

- *Sensitivity of test*—visual inspection is a traditional diagnostic method and it appears to have a very low sensitivity and high specificity in diagnosing caries.
- *Dry and clean teeth*—the teeth should be clean, dry and well illuminated during a visual examination to obtain maximum information.
- *Visual examination*—in detecting occlusal caries has a limited sensitivity. Black or brown discolorations and fissure morphology are not reliable for definitive

diagnosis of occlusal caries. The opacities of the enamel may be more useful in determining caries with visual examination as long as they are not stained but the teeth should be clean, dry and well illuminated.

Explorer

- *Sensitivity of test*—the sensitivity of the explorer is also reported to be low in diagnosing occlusal dentinal lesions. Use of sharp explorers in probing has been questioned by several authors. It is reported that it may cause damage or create a cavity at the site of a superficial carious lesion.
- *How to do it*—using explorer in the intraoral examination may not improve diagnostic accuracy. Sticking probe may not be indicating caries but only be a sign of local anatomical features.
- *Disadvantage*—also probing a sterile fissure after an infected one may inoculate pathogenic microorganisms and infect the sterile fissure. However, in a study which lasted as long as 11 years in which the same children were examined repeatedly as often as six times with vigorous probing of pits and fissures but it didn't show any evidence that probing increases caries. In some parts of the world, especially in Scandinavian countries, applying pressure with sharp explorer is not approved because of the damage, it would create change in the surface integrity and possible implantation of microorganisms. There are some controversial results in this issue but the evidence suggests that an explorer should be used lightly or not at all on occlusal surfaces. In most cases when there is a cavitation on the enamel surface, dentinal involvement is also there.

Radiographic Method

- *Bite wing radiography*—it is used for the diagnosis of proximal decay (**Fig. 21-1**) because caries tends to occur most frequently just below the contact point either mesially or distally.
- *Limitation of radiography in diagnosis of caries*
 - *Two dimensional image*—radiograph must be used with caution as it gives a two dimensional image of three dimensional objects. Due to this the exact site of the carious lesion cannot be located, i.e. buccal or lingual caries, the buccolingual extent of lesion, the distance between carious lesion and the pulp horns and the presence of recurrent caries in existing restoration may completely overlie the carious lesion.
 - *More mineral loss is required for radiographic detection*—another aspect is that the net mineral loss must exceed at least 20% to 30% in order to radiographically visible. Due to this, carious lesions are usually larger clinically than they appear radiographically and very early lesions are not evident at all on radiograph.

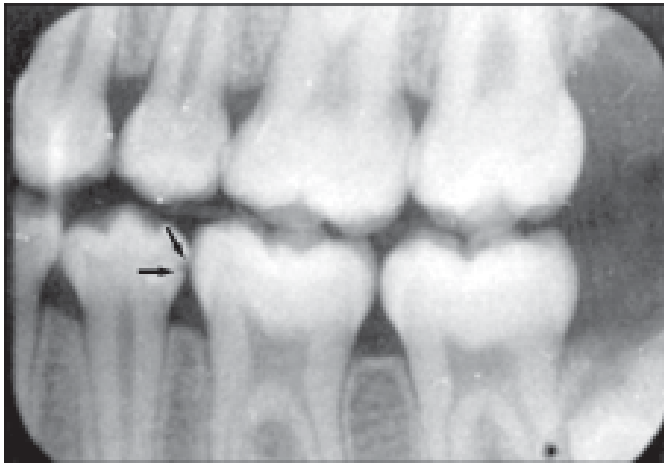


Fig. 21-1: Bite wing radiography is useful method for the diagnosis of dental proximal caries.

- *Technical variation*—technical variation in films and X-ray beam position can affect considerably the image of carious lesion, i.e. varying the horizontal tube head angulations can make lesion confined to enamel may appear to have progressed into dentin.

Fiberoptic Transilluminator (FOTI)

- *Sensitivity*—Fiberoptic transilluminator (FOTI) has been used since 1970s in the diagnosis of caries and it is a qualitative method.
- *Method*—a white light emitted from a cold light source is passed through a fiber to an intraoral fiberoptic light probe that is placed on the buccal or lingual side of the tooth. The surface of the tooth is examined using the transmitted light, seen from the occlusal view. Demineralized areas are darker when compared with the sound surrounding tissue. This contrast between sound and carious tissue is then used for detection of lesions.
- *Diagnosis of interproximal caries*—in the diagnosis of interproximal caries, fiberoptic transilluminator (FOTI) can also be used (**Fig. 21-2**). FOTI is reported to be superior to clinical examination and some researchers report that FOTI can detect 70%-90% of dentinal lesions. In terms of diagnostic accuracy and reliability, use of FOTI does not appear to provide any advantage over radiographs for diagnosing interproximal caries in clinical practice. It is difficult to estimate the depth of a lesion or the presence of a cavitation with conventional clinical examination as well as with FOTI in teeth with close interproximal contact.
- *Digital fiberoptic illuminator*—It is relatively new methodology that has developed in an attempt to reduce the shortcomings of FOTI, by combining FOTI and a digital camera. Illumination is delivered on the tooth

surface by means of fiberoptic illuminator which acts as a light source. The resultant change in light distribution is captured by the camera and is sent to the computer for analysis.



21-2: Detection of interproximal caries by using method of fiberoptic transillumination.

Electrical Conductance Measurement

- *Concept*—theory behind this is that sound surface should possess limited or no conductivity, whereas carious or demineralized enamel should have a measurable conductivity that will increase with increasing demineralization.
- *Results*—indicator for caries meter are four colored lights:
 - Green—no caries
 - Yellow—enamel caries
 - Orange—dentin caries
 - Red—pulpal involvement.

Visible Luminescent Spectroscopy

The visible emission spectrum for decayed and non-decayed regions of teeth differs. Quasi monochromatic light from a tungsten source dispersed with a grating monochromatic is focused on the teeth and emission spectra are recorded and analyzed.

Fluorescence

Acid dissolution of the structure results in a local decrease in fluorescence in area of acid exposure (**Fig. 21-3**). This has been used in detection of dental caries.

Use of Caries Detector Dye

Various dyes such as silver nitrate, methyl red and alizarin stain have been used to detect carious sites by change of color. The difficulty lies in removing the dye from the altered enamel area. The altered areas of enamel are characterized

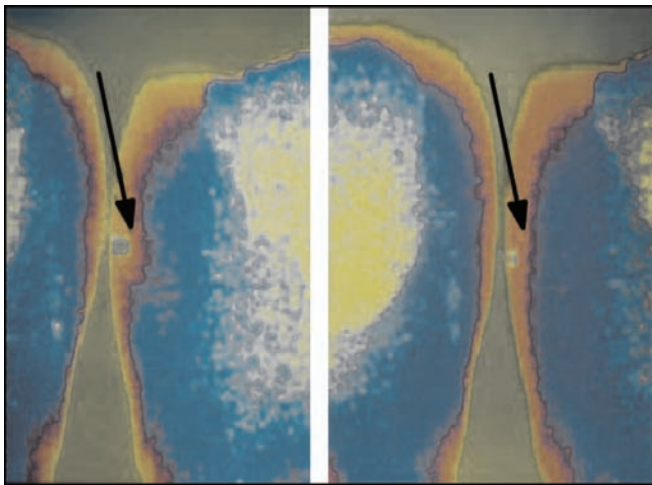


Fig. 21-3: Decreases fluorescence seen in proximal caries area

by more reactive calcium that reacts with carboxylic and sulfonic acid groups of dyes.

Types of Caries (Fig. 21-4)

Interproximal Caries

It is the type of smooth surface caries which is seen in interproximal area between the teeth (Fig. 21-5).

Clinical Features

- *Opaque chalky region*—it takes 3 to 4 years to manifest clinically as loss of enamel transparency resulting in opaque chalky region (white spot). In some cases, it

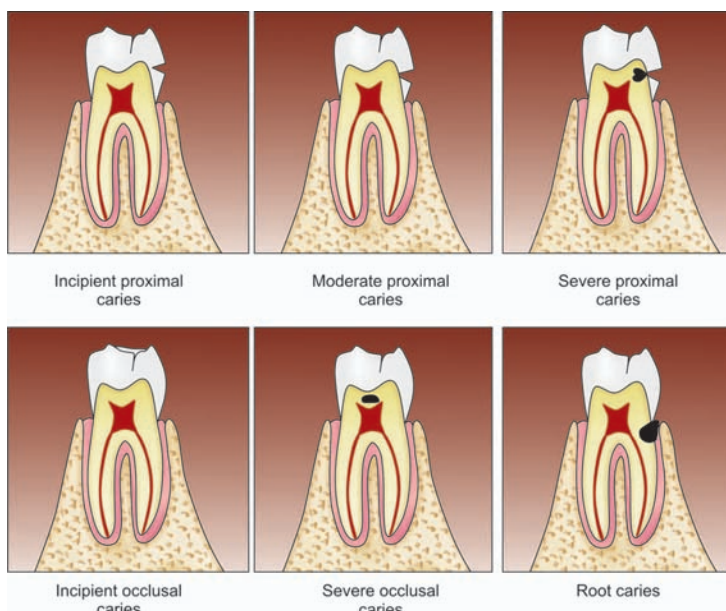


Fig. 21-4: Different types of carious lesion—a diagrammatic representation.



Fig. 21-5: Proximal caries seen with second premolar and first molar in proximal contact area.

appears as a yellow or brown pigmented area but it is usually well demarcated.

- *Location*—spots are generally located on the outer surface of enamel between contact point and height of free gingival margin. Caries do not initiate below free gingival margin.
- *Progress*—the early white chalky spot becomes slightly roughened owing to superficial decalcification of the enamel. As the caries penetrates the enamel, the enamel surrounding the lesion assumes bluish white appearance (Fig. 21-6) which is usually apparent as laterally spreading caries at the dentinoenamel junction. It is common for proximal caries to extend both buccally and lingually.
- *Pain*—as soon as carious process enter the dentin, patient complaint of sensitivity or pain with carious teeth.



Fig. 21-6: Interproximal carious lesion in anterior teeth showing bluish white appearance.

Radiographic Appearance

- *When it will detect*—radiographic detection of carious lesion on proximal surfaces of teeth depends on loss of enough material to result in detectable changes in radiographic density. As the proximal surfaces of posterior teeth are often broad, the loss of small amounts of mineral is difficult to diagnose on radiograph. 20 to 30% of demineralization is required for detection of lesion.

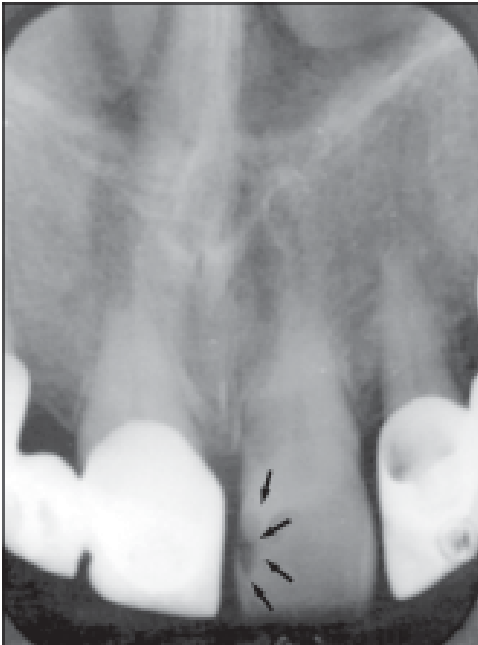


Fig. 21-7: Triangular shaped radiolucency seen with incisor tooth in proximal caries (Courtesy Dr Fusan Yasser).

- *Appearance*—interproximal carious lesions of enamel are triangular in shape (**Fig. 21-7**) and decrease in volume as they progress towards dentinoenamel junction. As the carious lesion approaches the DEJ, not only it become smaller but it is superimposed with more and more sound enamel which attenuates the X-ray and tends to obscure the demineralized lesion in proportion to its depth.
- *Fracture tooth*—in some cases due to severe caries tooth may get fracture (**Fig. 21-8**).

Radiological Types

- *Incipient caries*—radiographically, this caries susceptible zone has vertical dimension of 1.0 to 1.5 mm. There is loss of normal homogeneity of the enamel shadow (**Figs 21-9A and B**). It appears as a radiolucent notch on the outer surface of teeth. Magnifying glass should be used.
- *Moderate caries*—interproximal incipient lesion that develops and involves more than outer half of enamel but that do not radiographically extends into DEJ may

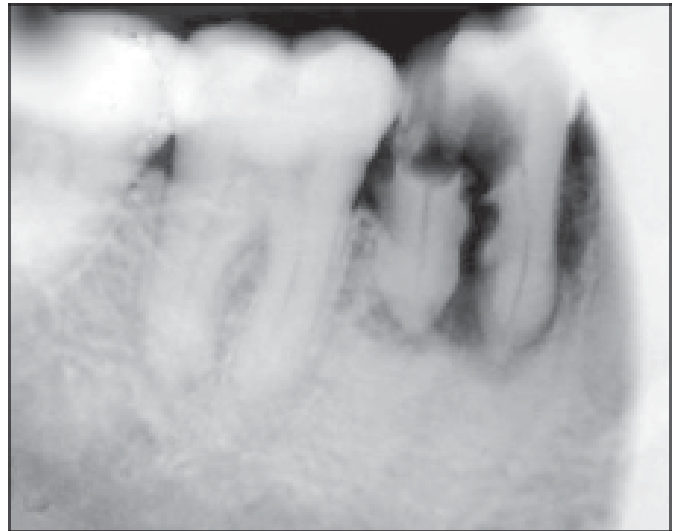
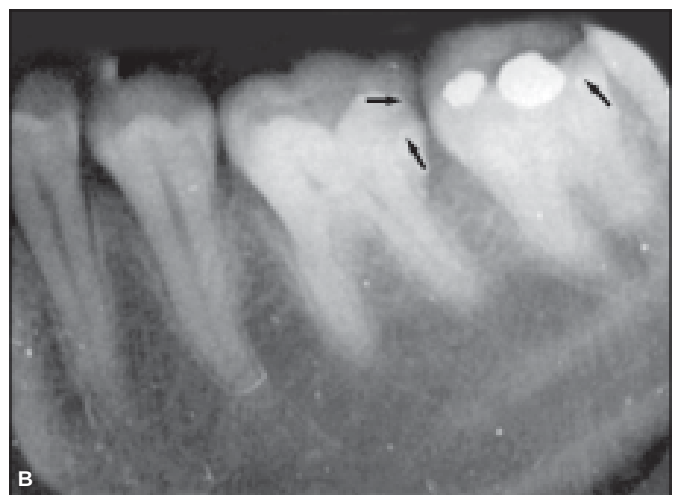
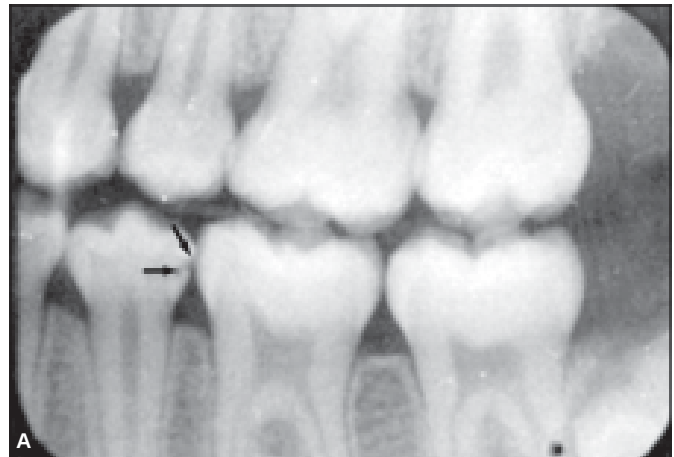


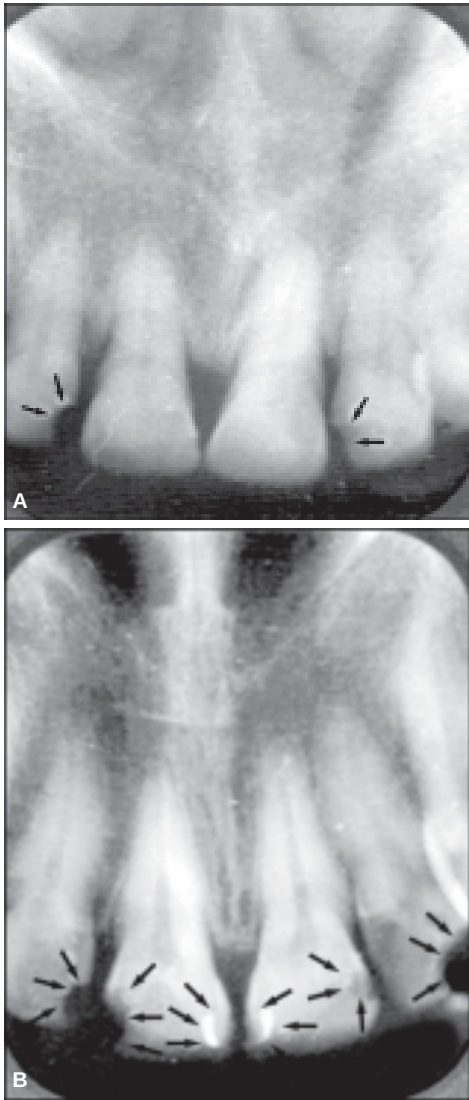
Fig. 21-8: Fractured tooth due to carious process.



Figs 21-9A and B: Incipient proximal caries showing loss of enamel homogeneity (Courtesy Dr Fusan Yasser).

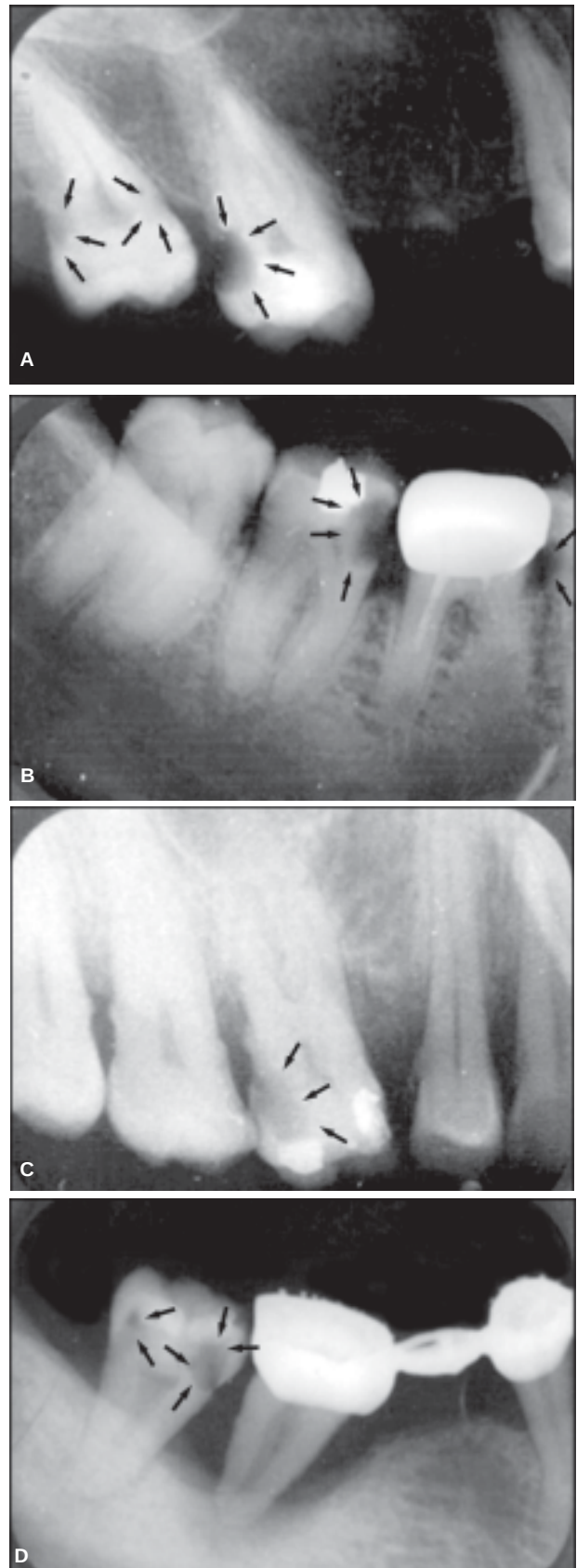
be called moderate lesion. Once the dentin is involved the margins of the radiolucent areas tapers off gradually into the adjacent tooth substance (**Figs 21-10A and B**). They have three radiographic appearances:

- Most common (67%) is that of triangle with broad base at the surface of tooth.
- Less common (16%) is diffuse radiolucent image.
- Third (17%) is a combination of the two types.

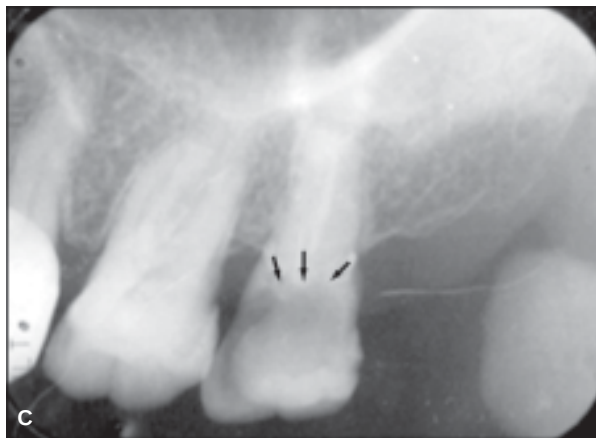
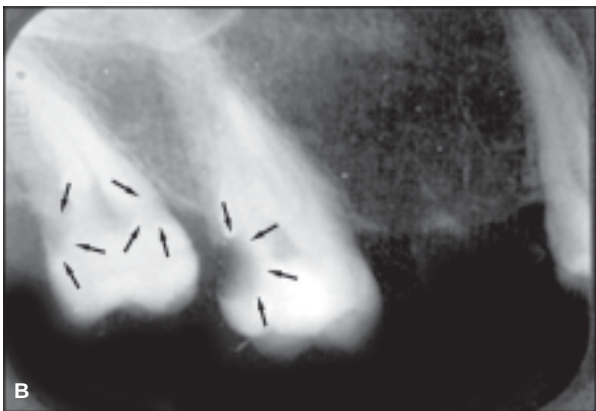
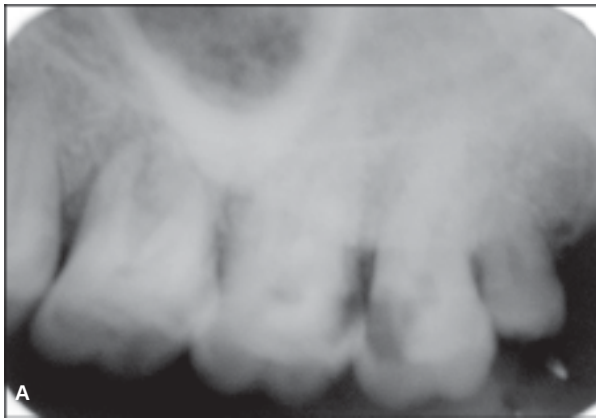


Figs 21-10A and B: Moderate caries seen in incisor region

- *Advanced caries*—these are the lesions that have invaded DEJ. Classically, there is more penetration through enamel. Configuration is usually triangular. It may be diffuse or combination of triangular and diffuse. Spreading of demineralization process at DEJ, undermining the enamel and subsequently extending into dentin which forms second irregular radiolucent image in dentin with base at DEJ and apex directed towards pulp (**Figs 21-11A to D**).



Figs 21-11A to D: Advanced caries showing extension in dentin (Courtesy Fusun Yasser),



Figs 21-12A to C: Severe carious lesion involving more than half of dentin (Courtesy Fusun Yasser).

- **Severe caries**—when carious lesion is seen radiographically to have penetrated through more than half of dentin (Figs 21-12A to C) and is approaching the pulp chamber, it is categorized as severe. It reveals a narrow path of destruction through enamel, the expansion of the radiolucency at DEJ and extends its development toward pulp chamber. It may or may not appear to involve pulp. Force of mastication will cause the undermined enamel to collapse leaving very large cavity or hole in the tooth (Fig. 21-12D).



Fig. 21-12D: Collapse of enamel occurs in severe proximal caries.

Cervical, Buccal, Lingual or Palatal Caries

There are many types of smooth surface caries occurring on cervical, buccal, lingual and palatal.

Clinical Features

- **Location**—it usually extends from the area opposite to the gingival crest occlusally to the convexity of the tooth surface. It extends laterally towards the proximal surfaces and on occasion extends beneath the free margin of the gingiva.
- **Cervical lesion**—it usually occurs in cervical area and the typical cervical lesion is a crescent shaped cavity (Fig. 21-13). Beginning as slightly roughened chalky area which gradually becomes excavated.
- **Buccal pit**—this again one of common type of caries which occur as pit on the buccal surface of tooth (Fig. 21.14).

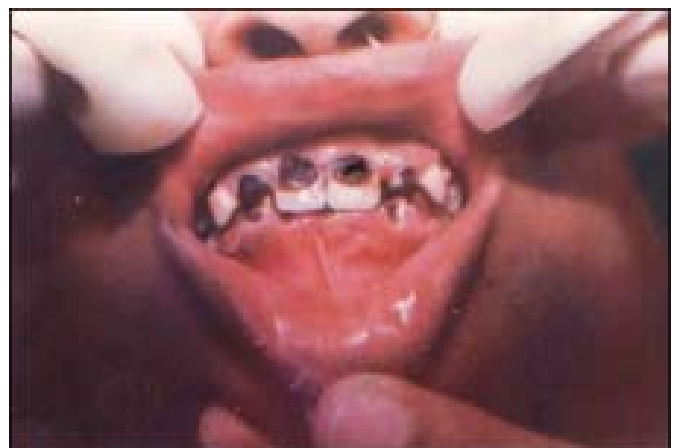


Fig. 21-13: Crescent shaped carious lesion of cervical caries.



Fig. 21-14: Buccal pit present on the buccal surface of tooth.

Radiographic Features

- It is difficult to differentiate between buccal and lingual caries on a radiograph.
- Appearance—there is uniform radiopaque circular region (Fig. 21-15) representing parallel non-carious enamel rod surrounding the buccal or palatal decay.

Pit and Fissure Caries

It is also called as 'occlusal caries'. It is primary type and develops in the occlusal surface of molars and premolars (Fig. 21-16). Deep narrow pits and fissures favor the retention of food debris and microorganisms and caries may result due to fermentation of food and the formation of acids.

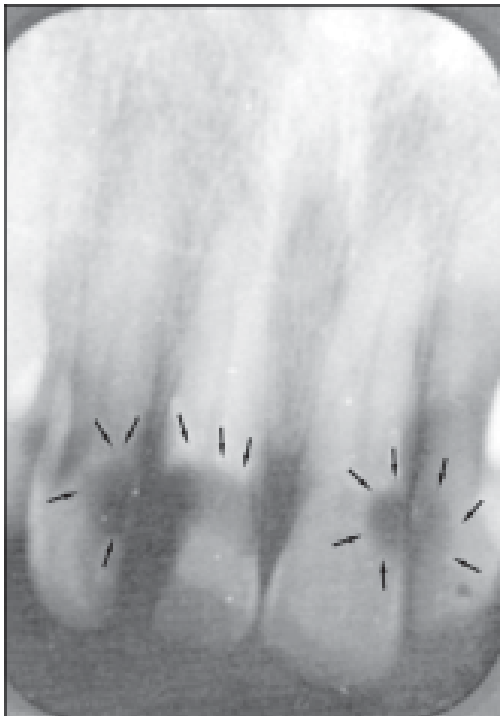


Fig. 21-15: Labial caries showing circular radiolucency.



Fig. 21-16: Deep pits and fissure more prone for carious tooth.

Clinical Features

- **Location**—it usually occurs in pits and fissures with high steep walls and narrow bases.
- **Appearance**—it appears brown or black (Figs 21-17A and B) and feels slightly soft and catches a fine explorer point. The enamel directly bordering the pit and fissure may appear opaque, bluish white as it becomes undermined.
- **Spread**—the lateral spread of caries at the dentinoenamel junction as well as penetration into the dentin along the dentinal tubules may be extensive without fracturing away the overhanging enamel. Thus, there may be large carious lesion with only a tiny point of opening.

Radiological Features

- **Common errors**—three common errors made in interpretation of occlusal caries are.
 - Failure to recognize that occlusal caries of enamel will not ordinarily be detected in the radiograph because of superimposition of heavy cuspal enamel over fissure area.
 - Carelessness in not observing rather long thin radiolucency that first appears at DEJ.
 - Confusion in distinguishing between occlusal and buccal caries.
- **Incipient lesion**—radiograph is not effective for the detection of an occlusal caries unless it reaches the dentin. The only change at the occlusal surface produced by early lesion is fine gray shadow just under the DEJ. Carious lesion generally starts at the side of fissure wall rather than its base. The lesion tends to penetrate nearly perpendicular to DEJ.
- **Moderate lesion**—there is broad base with thin radiolucent zone in the dentin with little or no changes in enamel (Fig. 21-18). There is band of increased opacity between carious lesion and pulp chamber.



Figs 21-17A and B: Pit and fissure carious lesion brown black discoloration of tooth.

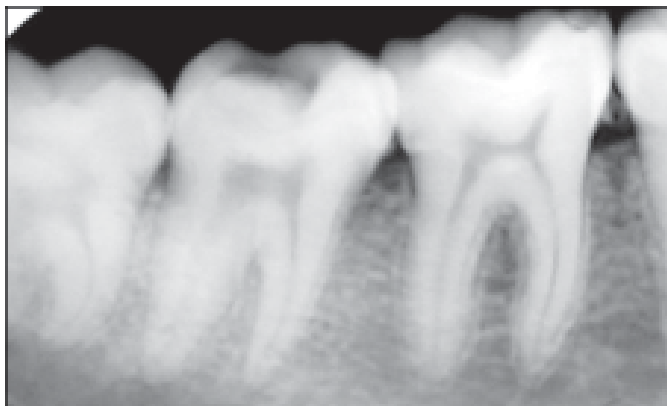
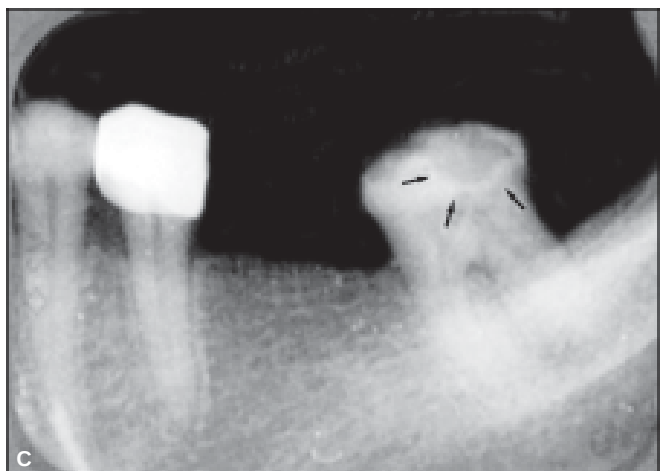


Fig. 21-18: Moderate occlusal carious lesion seen on 2nd molar.

- *Severe lesion*—large hole or cavity in the crown of the teeth (**Figs 21-19A to C**) Masticatory stress causes collapse of enamel.

Root Caries

It is also called as '*cemental caries*' and involves both dentin and cementum. Nowadays, there is greater prevalence of



Figs 21-19A to C: Severe occlusal carious lesion showing large hole in tooth.

root caries due to longer lifespan of persons, with the retention of teeth into the later decades of life and increase in the number of people exhibiting gingival recession with clinical exposure of cemental surface. Freshly exposed root are more vulnerable to an acid attack because of higher porosity and smaller crystal.

Clinical Features

- *Site*—it appears as slowly progressing chronic lesion. It is usually found in mandibular molar and premolar region. Tooth surface involved in decreasing order of frequency are buccal, lingual and interproximal.
- *Associated features*—gingival recession is associated with root surface caries (**Fig. 21-20**).



Fig. 21-20: Root surface caries seen in incisor region.

Radiographic Features

- *Appearance*—the carious process is best described as scooping out which result in radiographic appearance usually described as ill-defined saucer-like crater. If peripheral surface area is small, the appearance of carious lesion will be notched rather than saucer-like.

Recurrent Caries

Dental caries that occurs immediately adjacent to the restoration is referred to as recurrent caries (**Fig. 21-21**). It may be caused by inadequate extension of restoration and there has not been careful and complete excavation of original carious lesion.

Clinical Features

- *Incidence*—16% of restored teeth have recurrent caries.
- *Cause*—restoration will show poor margins which permit leakage and the entrance of both bacteria and substrate.



Fig. 21-21: Recurrent carious lesion seen below the restoration of tooth.

Radiographic Features

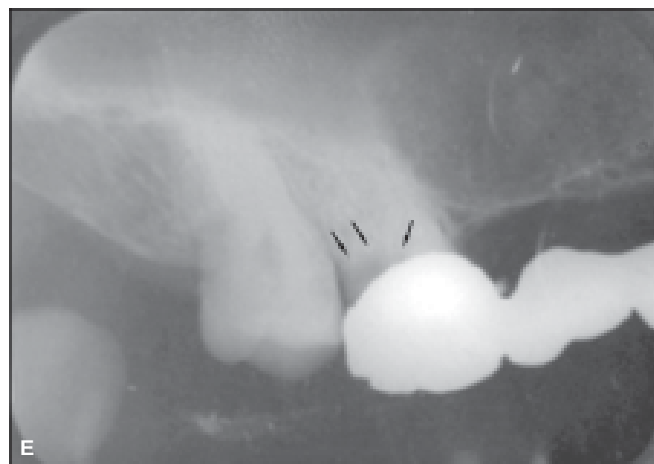
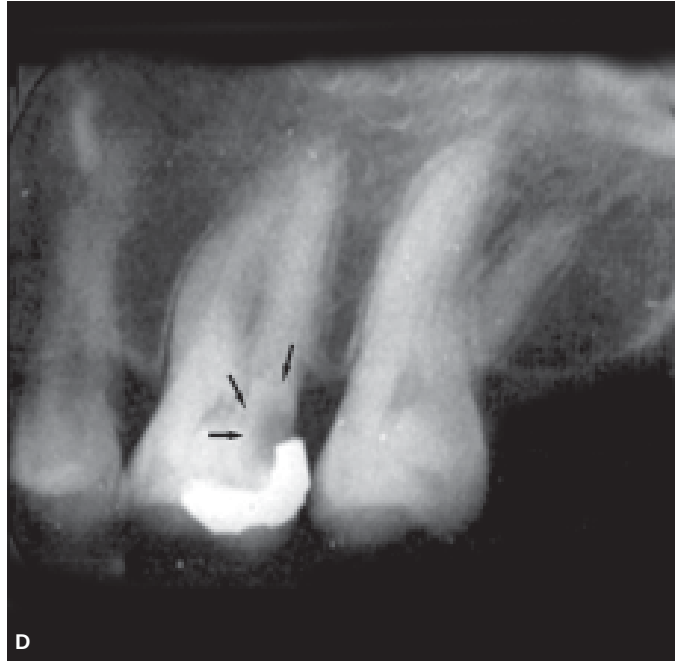
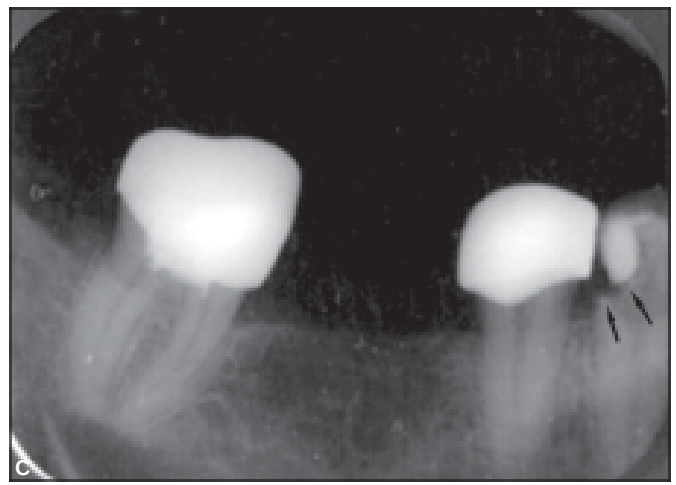
- *Appearance*—teeth have areas of increased radiolucency along the margins of the restoration. Recurrent caries that occurs at mesioingival, distoingival and occlusal margins are more frequently discovered than that which occurs at margins of buccal, facial and lingual restorations (**Figs 21-22A to E**).
- *Difficulty in diagnosis*—in dental amalgam mercury is mixed with an alloy powder containing silver, tin and zinc. With passage of time, tin and zinc ions are released into the underlying demineralized dentin producing a radiopaque zone within the dentin which follows the S-shaped curve of the underlying tubules. The radiopacities of this zone make the normal dentin on either side appear more radiolucent by contrast simulating recurrent caries and leading to difficulty in diagnosis.

Nursing Bottle Caries

This occurs in child who use nursing bottle in bed which contain milk or milk formulae, fruit juice or sweetened water. This type of caries is also prevalent in sugar or honey-sweetened pacifier.

Pathogenesis

The reasons for this are that the child is put on bed at afternoon nap time or at night with nursing bottle containing milk or a sugar containing beverage. The child fall asleep and the milk or sweetened liquid becomes pooled around the maxillary anterior teeth. The carbohydrate containing liquid provides an excellent culture medium for acidogenic microorganisms. Salivary flow is decreased during sleep and clearance of the liquid from the oral cavity is slowed. Lactose content of human milk, as well as that of



Figs 21-22A to E: Recurrent caries seen below the restoration (Courtesy Fusan Yasser).

bovine milk, can be cariogenic if the milk is allowed to stagnate on the teeth.

Clinical Features

- *Site*—there is early carious involvement of the maxillary anterior teeth (**Figs 21-23A and B**), the maxillary and mandibular first permanent molars, the mandibular canines.
- *Appearance*—the carious process in the teeth is so severe that only the root stumps remain.

Prevention

- *Holding the infant*—the infant should be held while feeding.
- *Remove the bottle from child mouth while he falls asleep*—the child who falls asleep while nursing should be burped and then placed in bed.



Figs 21-23A and B: Nursing bottle caries seen in maxillary anterior teeth (Courtesy Dr Shetty).

- *Brushing of child teeth*—parent should start brushing the child teeth as soon as they erupt in the oral cavity.
- *Discontinue bottle feeding after the age of 12 to 15 months*—discontinue bottle feeding as soon as child can drink from a cup, at approximately 12 to 15 months of age.

Radiation Caries

It is a rampant form of dental decay that may occur in individuals who receive a course of radiotherapy that includes exposure of the salivary glands.

Clinical Features

- *Appearance*—destruction begins at cervical region. The lesion may aggressively encircle the tooth causing the entire crown to be lost with only root fragment remaining in the jaws.
- *Clinical types*—clinically there are three types of radiation caries:
 - *Widespread superficial lesion*—it attacks buccal, occlusal, incisal and palatal surfaces.
 - *Circumferential caries*—it usually occurs in cementum and dentin in the cervical region. It may result in loss of the crown.
 - *Pigmentation of crown*—it is usually dark in color.

Radiological Features

- *Appearance*—radiographically it appears as dark radiolucent shadow appearing at necks of teeth most obvious on mesial and distal aspects.

Rampant Caries

It is defined as a suddenly appearing, widespread, rapidly burrowing type of caries, resulting in early involvement of the pulp and affecting those teeth usually regarded as immune to ordinary decay. Some believe that the term rampant caries should be applied to those carious lesions with 10 or more new lesions per year. It usually occurs in children with poor dietary habits.

Etiology

- *Systemic disease*—nutritional deficiency, malnutrition may lead to rampant caries.
- *Emotional disturbance*—emotional disturbances may be causative factors in some cases of rampant caries.
- *Stress and medication*—various forms of stress in both children and adults, as well as various medication (such as tranquilizers and sedatives) commonly taken to help persons cope with stress, are associated with decreased salivary flow and decreased caries resistance caused by impaired remineralization.

Clinical Features

- *Appearance*—it demonstrates extensive interproximal and smooth surface caries (**Figs 21-24 and 21-25**).
- *Occurrence*—rampant caries can occur suddenly in teeth that were for many years relatively immune to decay.



Fig. 21-24: Rampant form of caries showing total destruction of tooth surface in mandibular teeth.



Fig. 21-25: Rampant form of caries seen in mandibular teeth.

Radiographic Features

- *Appearance*—it demonstrate severe advanced carious lesions especially of mandibular anterior teeth.

Arrested Caries

It has been described as caries which becomes static or stationary and does not show any tendency for further progression.

Clinical Features

- *Location*—both deciduous and permanent dentitions are affected by this condition. It occurs exclusively in caries of occlusal surfaces.
- *Appearance*—it is characterized by large open cavities in which there is lack of food retention and in which the superficially softened and decalcified dentin is gradually burnished and has taken a brown-stained polished appearance and is hard. This has been referred to as 'eburnation of dentin'. In some cases, there is brown stained area at or just below the contact of the affected tooth.

Radiological Features

- *Appearance*—radiographically, these arrested carious lesions appear as small radiolucent areas (**Fig. 21-26**). Sometimes large occlusal carious lesions also arrest. In these lesions, there is an open cavity which is easily observed clinically.
- *Crown of tooth*—the crown of the tooth is absent and there is a radiolucent area in this region. Due to the defensive measure of the pulp to limit the carious lesion, there might be a sclerotic line under the radiolucent area.

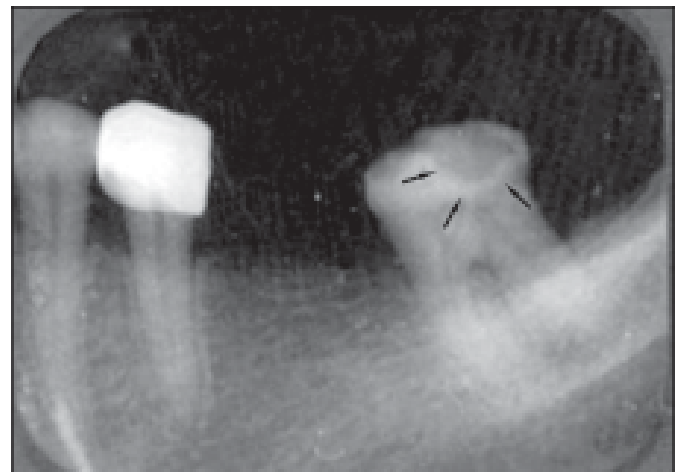


Fig. 21-26: Arrested caries showing small radiolucent area (Courtesy Dr Fusun Yasser).

Pre-eruptive Caries

Occasionally, defect on the crowns of developing permanent teeth are evident radiographically, even though no infection of the primary tooth or surrounding area is apparent. Such lesions resemble caries when it is observed clinically and the destructive nature of lesion progresses if it is not restored.

As soon as the lesion is reasonably accessible, the tooth should be uncovered by removal of the overlying primary tooth or by surgical exposure.

Linear Enamel Caries

This type of caries is seen in deciduous dentition in anterior maxillary teeth. Linear enamel caries is caused by metabolic disturbances or trauma. It is present in the region of neonatal line.

Acute Dental Caries

This type of caries runs rapid course and characterized by pulpal involvement in early course of caries. It is most commonly seen in young adults. The typical feature of acute dental caries is that, there is small opening, which will result in less contact of saliva with the carious process. This will result in less buffering and neutralization of acid. So the process will rapid. Pain is the constant feature of this disease.

Chronic Dental Caries

This is slow process and involvement of pulp occurs in a late stage. The opening of this carious process is large, resulting in less food retention. The dentin stained brown in this type of caries. There is little undermined enamel. Pain is not common feature of this disease.

Radiographic Differential Diagnosis of Dental Caries

- *Erosion cavity*—since these cavities are saucer shaped and have sloping margins in radiographs, they may resemble carious cavities. Clinical examination is necessary to rule out erosion cavity.
- *Non-opaque filling*—if carious cavities are filled with non-opaque filling material then it may resemble caries. But sharpness of the margin surgically prepared suggests the presence of filling.
- *Cervical burnout*—following is the difference between cervical burnout and cervical caries—
 - Cervical burnout is located at the neck of the teeth, demarcated above by the enamel cap or restoration and below by the alveolar bone level while caries have no apparent upper and lower demarcating borders.
 - It is triangular in shape, gradually becoming less apparent towards the center of the tooth while caries is saucer shaped and becomes more apparent toward center of tooth.
 - In cervical burnout axial border fades or follows anatomic contour while in case of cervical caries sharp delineation and ragged contour is present.
 - Peripheral outline of cervical burnout appear intact as compared to caries where it is cavitated.

- Usually all the teeth on the radiograph are affected, especially the smaller premolars while the caries is localized.
- *Internal resorption*—some of the cases will show pink tinge where the vascular pulp lies beneath the thin dental tissue. Margins of internal resorption are sharply defined as compared to caries which has fading margin due to decalcification. If the pulp chamber has its normal margin effaced, it is suggestive of internal resorption rather than caries.
- *External resorption*—only if the destruction is at the cervical portion of the tooth. External resorption may be mistaken for dental caries. In resorption, the line of demarcation between adjacent tooth substance and the defective area is sharp without any decalcification beyond the precise site of tooth destruction
- *Hypoplasia of enamel*—in hypoplasia, the radiolucent area is not single and it is common that several small dark spots cross the tooth. Visual inspection rule out the hypoplasia.

Control of Dental Caries

Control of all Active Lesions

Initial treatment of all active lesions should be done. Gross excavation of all carious lesions followed by restoring a tooth to normal contour is done.

Nutritional Measures for Caries Control

Group of patients whose diet is high in fat, low in carbohydrate and practically free from sugar have low caries activity. In a study, when refined sugar was added to the diet in the form of a mealtime supplement, there was little or no caries activity. Phosphates diet causes significant reduction in incidence of caries.

Mechanical Measures for Caries Control

- *Toothbrushing*—tooth brushing reduces the number of oral microorganisms, particularly if the teeth are brushed after each meal. Tooth brush also removes gross amounts of food debris and plaque material.
- *Mouth rinsing*—the use of mouthwash for the benefit of its action in loosening food debris from the teeth has been suggested as a measure of caries control.
- *Dental floss*—dental flossing has been shown to remove plaque from an area gingival to the contact areas on proximal surfaces of teeth, an area impossible to reach with toothbrush.

- *Detergent*—some workers have related the high caries incidence among modern civilized races to the unrestrained use of soft, sticky, refined foods, which tend to adhere to the teeth. It has been stated that fibrous food prevents lodging of food in pits and fissures of teeth and in addition acts as detergent.
- *Oral irrigators*—this is more useful in management of gingival infection.
- *Chewing gums*—this can prevent dental caries mechanical cleansing action.
- *Pit and fissure sealants*—pits and fissures of occlusal surface are among the most difficult areas on teeth to keep clean and from which to remove plaque. The pit and fissure sealants generally used in conjunction with an acid pretreatment to enhance their retention, contain either cyanoacrylate, polyurethane or the adduct of bisphenol A and glycidyl methacrylate.

Chemical Measures of Caries Control

- *Fluorine*—the cariostatic activity of fluoride involves several different mechanisms. The ingestion of fluoride results in its incorporation into the dentin and enamel of unerupted teeth. This makes the teeth more resistant to acid attack after eruption into oral cavity. In addition, ingested fluoride is secreted into saliva; although present in low concentration in saliva; the fluoride is accumulated in plaque where it decreases microbial acid production and enhances the remineralization of the underlying enamel. Fluoride from saliva is also incorporated into the enamel of newly erupted teeth, thereby enhancing the enamel calcification.
 - *Communal water fluoridation*—fluoridation of the communal water supply is the most effective method of reducing the dental caries problems in the general population.
 - *Fluoride containing dentifrices*—it contains stannous fluoride in combination with calcium pyrophosphate as the cleaning and polishing system and was accepted as the first therapeutic dentifrice.
 - *Fluoride mouth rinses*—it should be given cautiously in children under 4 years of age as they may not have full control over the swallowing reflex.
 - *Dietary fluoride supplement*—the administration of fluoride supplement commences shortly after birth and should continue through the time of eruption of the second permanent molars.
- *Bisbiguanides*—chlorhexidine and alexidine are potential anticaries agents as they are antiplaque agents. It has been shown that chlorhexidine is adsorbed onto tooth surface and salivary mucins then it is released slowly in an active form. But disadvantage of chlorhexidine is that it has bitter taste, produces brownish discoloration of hard and soft tissues and may produce painful desquamation of mucosa.
- *Silver nitrate*—silver nitrate impregnation of teeth was used for many years to prevent or arrest caries. Silver plugs the enamel by either the organic invasion pathways such as the enamel lamellae or the inorganic portion of enamel to form a less soluble combination.
- *Zinc chloride and potassium ferrocyanide*—use of solution of zinc chloride and potassium ferrocyanide would effectively impregnate the enamel and seal off caries invasion pathways. But study shows that it is of little value in reduction of caries.
- *Vitamin K*—synthetic vitamin K (2-methyl 1, 4 naphthoquinone) prevents acid formation in incubated mixture of glucose and saliva. In study also it has shown to decrease incidence of caries formation in persons given chewing gums containing vitamin K.
- *Sarcoside*—persons who brushed teeth with dentifrices containing sodium N-lauroyl sarcosinate have been shown to have decreased incidence of caries. It has been stated that it has got ability to penetrate the dental plaque and prevent pH to fall below 5.5 after carbohydrate rinse.
- *Urea and ammonium compounds*—reports suggest that a quinine urea mouthwash prevents acid formation in test *in vitro* on carbohydrate saliva mixtures. It may also be noted that oral bacteria count was decreased after the use of quinine urea mouthwash and salivary pH generally increased to value over 8 and remained high for approximately an hour. The evidence indicated that urea upon degradation by urease; release ammonia which act to neutralize the acids formed through carbohydrate digestion and also interferes with bacterial growth. Although there are some studies to indicate that ammoniated dentifrices are capable of producing some reduction in dental caries incidence, the magnitude of this reduction, particularly in persons whose tooth-brushing habits are not controlled or supervised, is not so great as to justify recommending them for widespread use as an anti-cariogenic agent.
- *Chlorophyll*—it is a green pigment of plants and has been proposed as an anti-cariogenic agent on the basis of a number of *in vitro* studies and animal studies. Water soluble form of chlorophyll, sodium copper chlorophyllin, was capable of preventing or reducing the pH fall in carbohydrate-saliva mixtures *in vitro*.
- *Nitrofurans*—they are derivatives of furfural which itself is derived from pentoses. They have been found to exert bacteriostatic and bactericidal action on many gram-positive and gram-negative bacteria and they can also inhibit acid formation. Studies show that nitrofurans compounds like furadroxyl (5-nitro-2-furaldehyde-2-hydroxyethyl semicarbazone) reduce dental caries.

- *Penicillin*—it has got ability to inhibit the normal biologic processes of lactobacilli which is one of the etiological factors in the dental caries. Penicillin is given in dentifrices.

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Introduction

The tongue makes up a large part of the oral cavity and can be affected by numerous lesions. The tongue may be affected as a part of oral disease or as a signs of a systemic disease. The word 'tongue' is derived from the Latin word '*lingua*' and Greek word '*glossa*'. It is partly oral (anterior 2/3rd of tongue) and partly pharyngeal (posterior 1/3rd of tongue). It is composed of body (movable oral part) and base (attached part).

Embryology and Development of Tongue

- *Origin*—the tongue develops from the ventral wall of the primitive oropharynx. It is derived principally from first, second and third branchial arch as well as from occipital myotomes.
- *Tuberculum impar*—tuberculum impar is the three centrally placed elevations on the ventromedial portion. But soon, elevations of the ventromedial portion of the 1st arch arise on each side of tubercle and merge with each other.
- *Foramen cecum*—there is transient elevation present on caudal border of tuberculum impar which is marked by a blind pit. This is called as foramen cecum.
- *Lingual swelling*—during the 4th week of development, paired lateral thickening of mesenchyme appears on the internal aspect of 1st branchial arch. This is called as lateral lingual swelling. Lateral lingual swelling merge with each other and overgrow the tuberculum impar to form the oral part of tongue.
- *Formation of anterior two-third of tongue*—anterior 2/3rd is formed by fusion of tuberculum impar and two lateral lingual swelling.

- *Formation of posterior 1/3rd of tongue*—the posterior 1/3rd of the tongue has a more complicated developmental origin. It first exists as a central mound called *the copula*, which is the result of fusion of the 3rd branchial arches. The endodermally derived mucosa of the 2nd to 4th branchial arches and the copula, provide covering for the posterior thirds of the tongue.
- *Sulcus terminalis*—it is the site of union between the base and the body of tongue. It is V shaped groove.
- *Occipital myotomes*—it migrates anteriorly into the tongue during fifth to seventh week. In a later stage various types of papillae differentiated in the dorsal mucosa of the body of tongue.
- *Tongue innervations*—ninth and twelfth nerves are carried along the migrating tongue mass. This mass then picks seventh and fifth nerve as it approaches the oral cavity.

Anatomy of Tongue

The tongue is a muscular organ situated in the floor of mouth, associated with the function of deglutition, taste and speech. Tongue has a base, body and a tip. It has two surfaces, a dorsal and a ventral surface. The dorsal surface is divided into an oral and pharyngeal part and the ventral surface is confined to the oral cavity only.

Surface

- *Sulcus terminalis*—it lies partly in the mouth (oral part), which comprises the anterior 2/3rd and in the pharynx (pharyngeal part), which comprises the posterior 1/3rd. Both the parts are separated by the inverted 'V' shaped sulcus called the *sulcus terminalis*. At the apex of sulcus terminalis, there is a depression, called *the foramen cecum*.
- *Anterior part*—in the anterior part of the tongue the mucous membrane is thin with reduced lamina propria

and is closely attached to the underlying muscular tissue. The color of the anterior part of the mucous membrane is pink and is marked by a variety of papillae that gives the tongue a characteristic roughness. The anterior part of the tongue is divided in half by the *median lingual sulcus*.

- *Posterior part*—the posterior part also called as *pharyngeal part* or base of the *tongue* is located posterior to the palatoglossal arch.
- *Lingual tonsil*—the surface without papillae shows a slightly corrugated appearance, due to the underlying lymphoid tissue called the *lingual tonsil*.
- *Root of tongue*—the root of the tongue is attached to the epiglottis by a medial fold (the glossoepiglottic fold). Laterally, pharyngoepiglottic (glossopharyngeal) folds pass from the sides of the tongue and pharyngeal wall to the epiglottis. The root of tongue is attached to the hyoid bone, below and the mandible above.
- *Ventral surface*—the ventral surface is smooth and purplish with no papillae. On the ventral surface, lingual veins are often visible as bluish streaks.
- *Lingual frenulum*—the tongue is connected to the floor of the mouth by a sickle shaped fold of mucous membrane called as *lingual frenulum*. Anteriorly, on either side of the frenulum, the *caruncles* opening for the submandibular ducts are visible.
- *Plica fimbriata*—at the lateral side of the vein, a fringed fold of mucous membrane called as the *plica fimbriata* or *fimbriated fold*.
- *Taste buds*—these are peripheral gustatory organs which are composed of modified epithelial cells. They are most numerous on the sides of circumvallate papillae and less on the walls surrounding the foliate papillae. They are more numerous in infants than in adults. With age, they undergo atrophy.

Papillae

- *Circumvallate papillae*—they are usually 8 to 12 in number and are the largest of the papillae. They are situated in a row parallel to and close to the sulcus terminalis. Papillae are 1 to 3 mm in diameter and are flattened with a circular depression. They are surrounded by a moat-like trough.
- *Fungiform papillae*—they are smaller than the vallate papillae and are distributed over the dorsal surface of the tongue, being most numerous on the anterior part. They are round and mushroom shaped and is distinguished from the filiform papillae by their larger size and bright red color. Their number is about 100/cm² on the tip and 50/cm² in the middle. They carry taste buds.
- *Filiform papillae*—these are smallest, but most numerous and are evenly distributed over the dorsum and are often

arranged in rows parallel to the sulcus terminalis, except for the tip where they run transversely. The papillae are conical, broadest at the base and whitish due to marked degree of keratinization. The concentration of papillae in man is calculated about 500/cm². They are more heavily concentrated in center of dorsum of tongue.

- *Foliate papillae*—they are vertical folds of the mucosa located at the margins of the tongue, just anteriorly to the palatoglossal arch.
- *Papillae simplices*—they are connective tissue papillae which are similar to the papillae of dermis of skin. They are present beneath the entire tongue surface, including the mucosal papillae described above.

Muscle

- *Types of muscle*—the muscles of tongue are grouped into two sets: an extrinsic set and an intrinsic set. The extrinsic muscles include genioglossus, hyoglossus, styloglossus and palatoglossus.
- *Genioglossus*—it is the largest and arises from the upper mental spine and spread in a fan-like fashion and is inserted into the tongue from its tip to the root. It draws the tongue forward and acting together. This muscle is able to flatten the tongue, making a concavity from side-to-side.
- *Hyoglossus*—it is a flat, quadrilateral muscle arising from the hyoid bone. It runs as thin plate upward, connect with fibers from the styloglossus and enter the tongue lateral to the genioglossus. It depresses the tongue.
- *Styloglossus*—it originates from the styloid process, passes downwards and forwards and inserts into the side of the tongue, connecting with fibers from the hyoglossus. The styloglossus draws the tongue upwards and backwards.
- *Palatoglossus*—it originates from the palate and runs in the palatoglossus arch, continuing into the side and dorsum of the tongue.
- *Intrinsic muscles*—these are situated inside the tongue and constitute a greater part of the organ. They are divided into the superior longitudinal, inferior longitudinal, transverse and the vertical muscles.
 - *Superior longitudinal*—it arise from submucous fibrous layer close to the epiglottis and from the median fibrous septum. It runs forward to the edges of tongue, some of its fibers being inserted into mucous membrane. It shortens the tongue and makes the dorsum concave, by turning the tip and side of the tongue upward.
 - *Inferior longitudinal*—it is a narrow band lying close to the inferior surface of tongue, between genioglossus and hyoglossus. It extends from the root apex of the tongue, some of its posterior fibers being connected with the body of hyoid bone. In front, it blends with the fibers of styloglossus. It shortens the tongue and

makes its dorsum convex by pulling the tip of tongue downwards.

- *Transverse muscle*—it originates from the median fibrous septum and runs in horizontal course, laterally to be inserted into submucous fibrous tissue. By their action, intrinsic muscles alter the shape of the tongue making it narrow and elongated.
- *Vertical muscle*—it is found at the borders of anterior part of tongue. Its fibers extend from dorsal to ventral surface, mainly near its lateral borders, but fibers are interspersed through the tongue. It makes the tongue broad and flattened.

Arterial Supply

- *Lingual artery*—the lingual artery, a branch of the external carotid, is the main vessel supplying the tongue. Before the artery reaches the posterior edge of the hyoglossus muscle, it gives off a branch to the hyoid bone area.
- *Lingual dorsal artery*—in its course below the hyoglossal muscle, it gives off a lingual dorsal artery, which runs steeply upward dividing into many branches supplying the base of tongue and posterior part of the dorsum.
- *Ascending pharyngeal artery and tonsillar artery*—the root of the tongue is also supplied by the ascending pharyngeal artery and tonsillar artery.
- *Sublingual artery*—at the lower border of the anterior part of the hyoglossal muscle, the lingual artery gives off the sublingual artery, which supplies the sublingual region medial to the sublingual gland.

Venous Drainage

- *Lingual vein*—deep lingual vein is the largest and the principal vein of the tongue. Deep lingual vein originates near the tip of the tongue and runs backward, close to the mucous membrane on the ventral surface of the tongue.
- *Vena comitans*—it joins the sublingual vein, at the posterior border of the hypoglossal muscle to form the *vena comitans* of the hypoglossal nerve, which drains to the facial or internal jugular vein.
- *Dorsal lingual vein*—the dorsal lingual vein drains into the dorsum and sides of the tongue. It joins the sublingual veins, which follow the artery deep to the hypoglossal muscle and enter the internal jugular vein, near the hyoid bone.

Nerve Supply

- *Hypoglossal nerve*—all the muscles of the tongue, except the palatoglossus are supplied by the *hypoglossal nerve*. The palatoglossus is supplied by the pharyngeal plexus.
- *Lingual branch* of mandibular nerve is nerve for general sensation for anterior 2/3rd of tongue.

- *Glossopharyngeal nerve* is the nerve for general sensation for posterior 1/3rd of the tongue.
- Posterior most part of tongue is supplied by vagus nerve, through *internal laryngeal nerve*.
- Taste sensation is carried out by *chorda tympani* branch of facial nerve for anterior 2/3rd and glossopharyngeal nerve for posterior 1/3rd.

Lymphatic Drainage

- *Submental nodes*—the tip of the tongue drains bilaterally into the submental nodes.
- *Submandibular lymph nodes*—right and left halves of rest of the anterior 2/3rd of the tongue, drain unilaterally to submandibular lymph nodes. A few central lymphs drain laterally into the same nodes.
- *Jugulodigastric nodes*—some of the lymph vessels, from the lateral margins of the tongue, drain to the jugulodigastric nodes.
- *Jugulo-omohyoid nodes*—the posterior 1/3rd of the tongue drain bilaterally to the jugulo-omohyoid nodes, in which most of the lymphs drain from the tongue.

Functions of Tongue

- *Speech*—it is the result of interaction between different organs. Even small changes in the position or shape of the tongue may cause disturbance in speech. Tongue is one of the organs in the oral cavity, which interrupts the air passage through mouth or pharynx thereby producing consonants. Certain consonants like c, d, j, i, n, t, z, l, g, etc. require movement of tongue.
- *Mastication*—the tongue has a direct crushing effect on food by pressing it against the hard palate. The tongue pushes the food onto the occluding surfaces and helps to mix in the saliva. The sensory ending on the tongue enable to select those parts of the food mass are sufficiently well masticated to be ready for swallowing.
- *Deglutition*—when the food bolus is placed on dorsum of tongue, it is pressed lightly against the hard palate just behind the incisors. It is a coordinated muscular activity involving the tongue and constrictor muscle of the pharynx, to close the palatal vault and the epiglottis. It allows the passage of the bolus into the esophagus, without regurgitation into the nose or lower respiratory tract. The process is initiated by the voluntary action of collecting food onto the tongue and propelling it backwards into the pharynx. The muscles involved in this process are the mylohyoid and the pharyngeal constrictors. Bolus is pushed backwards by raising the back of tongue. Food bolus is sucked from mouth into pharynx, by creating a negative pressure, while airways

are still closed by rapid relaxation of muscles of tongue and pharynx.

- *Digestion*—tongue has a slight digestive function by virtue of salivary lipase, present in serous lingual salivary glands.
- *Taste*—it acts as a special sense organ of taste, by virtue of presence of numerous taste buds. The tip of the tongue is most sensitive to substances eliciting a sweet sensation. The lateral margins are most sensitive to substances causing sour sensation. The base of the tongue is most sensitive to substances eliciting a bitter sensation. The salty quality is more widespread, but is greatest at the tip.
- *Barrier function*—mucosa covering the tongue acts as a barrier protecting the deeper tissues from mechanical damage. It also prevents entry of microorganisms and toxic substances.
- *Jaw development*—muscular pressure from the tongue is an important factor in determining the shape of the mandibular arch.
- *Thermal regulation*—it is more pronounced in dogs, where there is a considerable loss of heat from the tongue.
- *Secretion*—major secretion of tongue is provided by salivary glands activity which maintains the moist surface of oral mucosa.
- *Defense mechanism*—secretory immunoglobulin system of tongue plays an important role in body defense.
- *Maintenance of oral hygiene*—by virtue of its movement it can reach all parts of the oral cavity removing food debris from the gums, vestibule and floor of mouth. Thus, it helps in maintenance of oral hygiene.
- *Sucking*—tongue also plays an important role in sucking in both bottle feeding and breastfeeding.
- *General sensitivity*—due to extreme sensory innervations terminating in both simple and organized nerve endings, there is perception of heat, cold, pressure and chemical discrimination.
- *Symbolic function*—functions that are traditionally associated with the tongue, but that have no anatomic and physiologic basis should be mentioned because images of this type are well established, cultural and literary tradition. It must frequently influences a patient perception of a lingual abnormality. Expressions such as 'speaking with a forked tongue' or 'speaking in different tongue' all describe the mental attitude and behaviors, by which they are expressed.

Specialized Examination of the Tongue

- *Cineradiography*—cineradiographic studies of the oral cavity and pharynx during drinking, chewing, suckling,

phonation and other activity, have added immeasurable to our understanding of the position and shape of the tongue in motion. It also helps in the diagnosis of abnormalities of swallowing, phonation and other functions, associated with congenital and surgically induced defects.

- *Computer assisted tomography*—it is used for number of instances to identify the space occupying lesion and muscular atrophy secondary to hypoglossal nerve damage, where the lesion was deep in the base of the tongue.
- *Pulsed (Doppler) ultrasound*—it is used to study the characteristic of arterial blood flow in the tongue and abnormal pulse wave in the lingual artery of individuals with evidence of compromise flow in other branches of carotid artery.
- *Real time ultrasound*—in it, probes of sufficiently small cross sectional diameter are used for exploring the ventral surface of the tongue. It is used to produce image of a cyst or other lesion within the tongue and also to estimate tongue size.
- *Isotopic scanning technique*—it is useful where the mass in the tongue is composed of specialized secretory tissue or other tissue, such as thyroid, which selectively concentrates intravenously administered radioactive elements.
- *Electromyography*—it is used to study the action potential in actively contracting muscles. It has contributed to an understanding of lingual and masticatory muscular function.
- *Scanning electron microscope*—it is useful for studying the surface topography of the tongue dorsum and the character and morphology of different types of tongue papillae.
- *Transmission electron microscopy*—it is used to study the pathologic changes in the taste buds in xerostomia and lesions of the 7th and 9th cranial nerves.
- *Taste testing*—the midline of the tongue and the V-shaped row of papillae separates the tongue into four quadrants. The anterior two of which bear fungiform papillae, with gustatory receptors connected via the right and left lingual branches of the 5th cranial nerve to the chorda tympani and facial nerve. The posterior two quadrants include the right and left vallates and the pharyngeal surface of the tongue, innervated by gustatory fibers of the lingual branches of the right and left glosso-pharyngeal and possibly, the vagus nerve. Testing of these four quadrants, thus, allows separation of abnormalities of the right and left side and 5th versus 9th cranial nerve function. Tongue must be dried or rinsed between each application of testant and it must be held in extruded position from the time of each

Table 22-1: Classification of tongue disorders

Developmental		
• Aglossia or hypoglossia	• Adenoma	• Syphilitic mucus patches
• Ankyloglossia	• Hemangioma	• Verruca vulgaris
• Bifid tongue	• Lymphangioma	
• Lingual polyp		Neurologic
• Macroglossia	Malignant	• Dyskinesia—involuntary movements
• Midline fistula	• Squamous cell carcinoma	• Glossodynia
• Teratoma	• Adenocarcinoma	• Trigeminal neuralgia
• Median rhomboid glossitis	• Transitional cell carcinoma	• Glossopharyngeal neuralgia
	• Verrucous carcinoma	• Polyneuritis
	• Mucoepidermoid carcinoma	• Neurofibromatosis
	• Reticular cell carcinoma	• Tongue thrusting
Infectious		• Dysgeusia
• Bacterial	Metastatic lesions from	
• Fungal and saprophytic	• Kidney	Papillary changes in tongue
• Parasitic	• Liver	Atrophic
• Viral	• Stomach	• Median rhomboid glossitis
	• Lung	• Geographic tongue
Cystic		• Pernicious anemia
• Epidermoid	Fissured tongue	• Protein deficiency
• Dermoid	• Congenital	• Lichen planus
• Lymphoepithelial	• Syphilis	• OSMF
• Mucus	• Amyloidosis	• Scleroderma
• Anterior median lingual cyst	• Melkersson-Rosenthal syndrome	
• Gastric mucosal cyst	• Papillon-Lefevre syndrome	Hypertrophic
• Parasitic cyst	• Traumatic bite	• White and black hairy tongue
• Bronchogenic cyst	• Lymphosarcoma	• After antibiotic therapy
	• Angiosarcoma	• After steroid therapy
	• Kaposi's sarcoma	• Hydrogen peroxide mouth wash
Neoplastic	• Melanoma	• Immunosuppressive drugs
Benign	Red and white lesions	• Smoking
• Fibroma	• Leukoplakia	• High fever
• Granular cell myoblastoma	• Erythroplakia	• Constipation
• Glomus tumor	• Lichen planus	• Hyperacidity
• Leiomyoma	• OSMF	
• Rhabdomyoma	• Candidiasis	Systemic diseases manifested in tongue
• Rhabdomyosarcoma	• Psoriasis	• Infections—bacterial, viral and fungal
• Neurofibroma	• Focal epithelial hyperplasia	• Blood disorders
• Keratoacanthoma	• White sponge nevus	• Metabolic disorders
• Traumatic neuroma	• Pemphigus	• Dermatological disorders
• Papilloma		• Collagen and autoimmune disorders

application until the patient gives his response. The drops of solution are best applied with the help of a pipette, rather than a cotton swab. The patient should be allowed to rinse his mouth with tap water, from time to time and to retract his tongue and relax between tastes.

Classification of Tongue Disorders

Classification of tongue disorders is discussed in Tables 22-1 to 22-3.

Congenital and Developmental Disorders

Aglossia and Microglossia

Definition

- *Aglossia*—it is the complete absent of tongue at birth.
- *Microglossia*—it is the presence of small rudimentary tongue.

Clinical Features

- *Symptoms*—patient encounters difficulty in eating and speaking.
- *Signs*—patient may have high arched palate and a narrow constricted mandible.
- *Airway problems*—there may be an airway obstruction, due to negative pressure generated by deglutition and inspiration.
- *Syndrome associated*—oromandibular limb hypogenesis syndrome (hypodactylia) and hypomelia and Pierre Robin syndrome.

Diagnosis

It can diagnose on clinical examination.

Management

- Non-surgical technique such as positioning, nasogastric intubation and temporary endotracheal intubation can be carried out to prevent airway obstruction.

Table 22-2: Classification of tongue disorders

Inherited, congenital and developmental anomalies**Minor variations**

- Partial ankyloglossia
- Variation in tongue movement
- Tongue thrusting
- Fissured tongue
- Patent thyroglossal duct and cyst
- Lingual thyroid
- Median rhomboidal glossitis

Major variations

- Cleft, lobed, bifurcated and tetrafurcated tongue
- Aglossia, hypoglossia and macroglossia
- Hamartoma and dermoid
- Bald and depapillated tongue
- Papillomatous changes

Disorders of lingual mucosa**Changes in tongue papillae**

- Geographic tongue
- Coated or hairy tongue
- Neuromuscular disorders
- Sleep apnea syndrome
- MPDS
- Vascular disorders

Non-keratotic lesion

- Thrush
- White sponge nevus
- Vesiculobullous and other desquamative disorders

Keratotic white lesions

- Lichen planus
- Leukoplakia

Depapillation and atrophic lesions

- Chronic trauma
- Nutritional deficiency
- Hematological abnormalities
- Medication
- Peripheral vascular disease
- Diabetes and chronic candidiasis
- Tertiary syphilis and interstitial glossitis

Pigmentation

- Ulcer and infectious disease
- Superficial vascular disease

Disorders affecting body of mandible

- Amyloidosis
- Infection

Tumors of tongue

- Squamous cell carcinoma
- Benign

Table 22-3: Classification of tongue disorders

Congenital and developmental disorders

- Aglossia and microglossia
- Macroglossia
- Ankyloglossia
- Cleft tongue
- Ankyloglossum superius syndrome
- Lingual varices
- Lingual thyroid nodule
- Variations in tongue movement
- Patent thyroglossal duct cyst
- Tongue thrusting
- Lingual polyp
- Reactive lymphoid aggregate
- Lingual cyst

Local tongue disorders

- Fissured tongue
- Median rhomboidal glossitis
- Benign migratory glossitis
- Hairy tongue
- Crenated tongue
- Foliate papillitis
- Leukokeratosis nicotine glossitis

Depapillation of tongue**Local disease**

- Eosinophilic granuloma
- Traumatic injuries
- Lesions due to automutilation
- Allergic stomatitis
- Facial hemiatrophy
- Cranial arteritis
- Chronic candidiasis

Systemic disease

- Iron deficiency anemia
- Plummer-Vinson's syndrome
- Pernicious anemia
- Niacin deficiency
- Folic acid deficiency
- Peripheral vascular disease

- Dermatomyositis
- Diabetes
- Syphilis
- Zoster infection
- Tuberculosis

Neurological disease

- Glossodynia
- Dyskinesia
- Paralysis
- Oropharyngeal dysphagia

Cyst

- Anterior median lingual cyst
- Bronchogenic cyst
- Epidermoid and dermoid cyst
- Gastric mucosal cyst
- Parasitic cyst
- Thyroglossal duct cyst

Benign tumor

- Fibroma
- Glomus tumor
- Granular cell tumor
- Leiomyoma
- Rhabdomyoma
- Plasmacytoma

Pre-malignant lesion and conditions

- Leukoplakia
- Lichen planus
- Oral submucous fibrosis

Malignant tumor

- Squamous cell carcinoma
- Malignant lymphoma
- Malignant melanoma
- Metastatic tumor
- Sarcoma

Miscellaneous

- Pigmentation of tongue
- Phlebotasia

Macroglossia

Macroglossia is tongue enlargement, which leads to functional and cosmetic problems. Although, this is relatively uncommon disorder, it may cause significant morbidity. Normal speech and swallowing reflexes require normal tongue anatomy and its functions. Swallowing begins as the tongue mixes food with saliva to form a food bolus, which is then propelled into the pharynx by the tongue. Articulation also depends on the tongue's ability to alter the impedance of air and change the resonant characteristics of the upper airway. In macroglossia, increased tongue bulk may impair these functions.

Classification

- *Congenital*
- *Acquired*
 - *Hypertrophic*—in it, muscles of the tongue are hypertrophic. It usually occurs in mentally retarded patients.
 - *Inflammatory*—it may involve the tongue partially or completely. It is due to various causes like syphilitic, Ludwig's angina, etc.
 - *Neoplastic*—it can be based on benign and malignant tumors.
- *Relative macroglossia*—it is a condition, in which a normal sized tongue appears abnormally large, if it is particularly enclosed within a small oral cavity.
- *Apparent macroglossia*—it is a condition where the tongue appears large due to poor muscular control of the tongue, although there is no increase in the bulk of tongue tissue.

Etiology

- *Congenital*—it includes hemangioma, lymphangioma and lingual thyroid. Other congenital disorders which can cause macroglossia are cretinism, Down syndrome, neurofibromatosis and multiple endocrine neoplasia type 2B
- *Inflammatory*—inflammatory causes include tuberculosis, actinomycosis, dental infection, syphilitic gumma, Riga disease, ranula and sublingual calculus.
- *Traumatic*—traumatic causes include dental irritation, hematoma and postoperative edema.
- *Neoplastic*—the neoplastic causes can be divided into malignant and benign lesions; with the malignant lesions including carcinoma and sarcoma. The benign lesions include granular cell tumor, neurofibroma, leiomyoma and lipoma.
- *Metabolic*—metabolic causes are myxedema, amyloidosis, lipid proteinosis, chronic steroid therapy and acromegaly.
- *Muscular hypertrophy*—over development of musculature, this may or may not be associated with generalized muscular hypertrophy or hemihypertrophy.

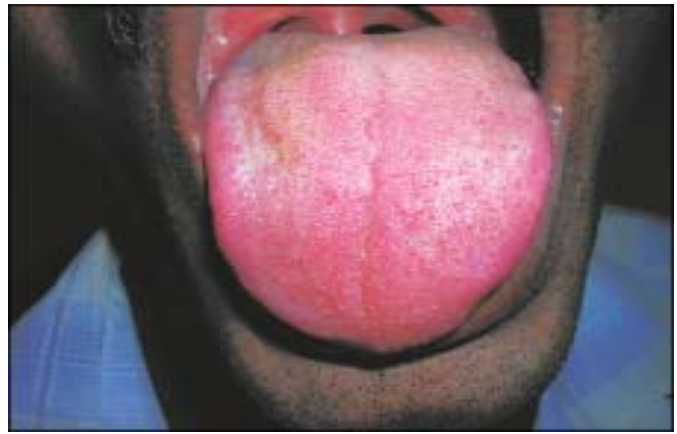


Fig. 22-1: Macroglossia showing protruded tongue.

Clinical Features

- *Age*—macroglossia is most prominent in infants, but tongue size may remain above normal in childhood and adolescence. As hyoid descends with age, macroglossia may improve.
- *Symptoms*—patient complaint of noisy breathing, drooling of saliva and difficulty in eating. Patient may get recurrent upper respiratory tract infection as tongue is usually protruded (Fig. 22-1) and mucosal drying occurs. The enlargement is generalized and may cause variety of difficulties with speech, feeding and airway problems.
- *Signs*—It may produce displacement of teeth and malocclusion, due to the strength of muscles involved and pressure exerted by the tongue on teeth.
- *Crenated lateral border*—crenation or scalloping of the lateral borders of the tongue; the tips of scalloping fit into the interproximal spaces between the teeth.
- *Syndrome associated*—it is associated with syndromes like Beckwith's hypoglycemic syndrome which includes neonatal hypoglycemia, mild microcephaly, umbilical hernia, and fetal visceromegaly and postnatal somatic gigantism.

Diagnosis

- *Clinical diagnosis*—large size of the tongue can be clinically diagnosed.

Management

- *Orofacial therapy*—it uses a palatal device to stimulate muscular tone and proper tongue position.
- *Surgical management*—majority of the cases of macroglossia are treated surgically. Indications for surgery include airway obstruction, speech difficulties, dysphagia and cosmetics. The procedure of choice is partial glossectomy. Surgical goal is to reduce the tongue size and thus improve the condition.

- *Removal of cause*—removal of primary cause should be done.
- *Orthodontic treatment*—correction of the dental arch deformity and malocclusion by orthodontic treatment.
- *Speech therapy*—correction of defective articulation by speech therapy.

Ankyloglossia

It is also called as 'tongue-tie'. It occurs due to incomplete degeneration of cells while the body of tongue is freed. In it, tip of tongue remains tied to floor of mouth. It is a condition in which the lingual frenulum is either too short or anteriorly placed limiting the mobility of the tongue. Reports of partial ankyloglossia affecting several generations, suggest a possible genetic basis for the minor variation in the attachment of the genioglossus muscle. It may be traumatic or congenital.

Types

- *Complete*—fusion of tongue and the floor of mouth.
- *Partial*—short lingual frenum.

Clinical Features

- *Symptoms*
 - *Restricted tongue movement*—it may limit the movement of the tongue.
 - *Feeding problems*—In extreme cases of ankyloglossia, nursing and feeding problems can occur. Poor sucking and inability to chew some food also occurs.
 - *Speech defect*—it was felt that tongue-tie was associated with speech abnormalities, especially lisping and inability to pronounce certain sounds and words viz t, d, n, l, as, ta, te, time etc.
 - *Tongue biting*—in some cases, recurrent tongue biting is also reported.
- *Signs*
 - *V shaped notch*—when there is an attempt to stick the tongue out, there may be a V shaped notch at the tip (Fig. 22-2). Physical examination will easily demonstrate the short or anteriorly placed lingual frenulum (Fig. 22-3).
 - *Anterior open bite*—patients have midline mandibular diastema and inability to clean the teeth and lick the lips with tongue.
 - *Periodontal problems*—due to high frenum attachment some patient may face periodontal problems.
- *Syndromes associated*—ankyloglossum superius syndrome, Rainbow syndrome, Fraser's syndrome and orofacial digital syndrome.

Diagnosis

- *Clinical diagnosis*—it can be easily diagnosed clinically.



Fig. 22-2: V shaped notch seen at the tip of tongue in patient with tongue-tie.



Fig. 22-3: Anteriorly placed lingual frenum seen in patient of tongue-tie (Courtesy Dr Chole).

Management

- *Counselling*—physician education, parental education and reassurance should be given to the patient.
- *Surgery*—indications for surgery, i.e. frenectomy are as follows:
 - If complete fusion of tongue is present then it requires surgery.
 - When nursing and feeding become a problem, surgery is indicated.
 - Children between 2-4 years, with poor development of speech and anxious parent's desire for the necessary treatment.
 - In cases where tongue-tie has recurred after snipping.
- *Complications of surgery*
 - Injudicious cutting of the frenum in a newborn can cause hemorrhage and the tongue may become too mobile and may be swallowed, causing asphyxia.

- There is also possibility of a subsequent infection at the base of the tongue, with formation of large ulcer and spreading stomatitis.

Cleft Tongue

It is also called as '*bifid tongue*'. It is the condition in which there is cleavage of the tongue due to lack of fusion of the lateral halves.

Pathogenesis

- *Failure of merging*—there is failure of merging of two lateral lingual swelling. Underlying mesenchymal proliferation fails to obliterate the groove. As these masses grow anteriorly, they move apart.

Clinical Features

- *Incidence*—completely bifid or cleft tongue is rare. It is due to lack of merging of lateral swellings of the organ.
- *Appearance*—partially cleft tongue is manifested as deep grooves in the midline of dorsal surface (**Fig. 22-4**).
- *Symptoms*—food debris and microorganisms may collect in the base of cleft and cause irritation.
- *Syndromes associated*—seen with orofacial-digital syndrome, with thick fibrous bands in lower anterior mucobuccal fold, which eliminate the sulcus and is associated with clefting of hypoplastic mandibular alveolar process.



Fig. 22-4: Partially cleft tongue manifested as deep groove in the tongue.

Diagnosis

- *Clinical diagnosis*—cleft can be seen clinically.

Management

- Regular cleaning of the tongue should be carried out.

Ankyloglossum Superius Syndrome

It is characterized by the attachment of the tongue to the hard palate and by limb malformation. It is also caused by genetic and intrauterine environmental factors.

It is a very rare disorder and is associated with jejunal and ileal atresias. Patient has difficulty in speech and mastication. Patient may have difficulty in swallowing. Surgical separation of the tongue from the palate.

Lingual Varices

A varix is a dilated, tortuous vessel (**Fig. 22-5**), most commonly a vein, which is subjected to increased hydrostatic pressure and is poorly supported by the surrounding tissues. Varices involving the lingual veins are relatively common, appearing as red or purple shot-like clusters of vessels on the ventral surface and lateral borders of the tongue as well as in the floor of the mouth. There appears to be no significant relationship between the presence of leg varicosities and sublingual varices.



Fig. 22-5: Tortuous vessels seen on ventral surface of tongue.

Lingual Thyroid Nodule

Presence of thyroid tissue in any region other than normal is called as ectopic thyroid gland. It is rare congenital anomaly which results from failure of descent of the primitive thyroid from the foramina caecum.

Pathogenesis

- *Development of thyroid gland*—the thyroid gland develops in the embryo from the ventral floor of the pharynx, by means of an ectodermal invagination of diverticulum. The thyroid gland tissues are formed at the median end of the two mandibular arches which are separated by tuberculum impar.

- *Development of tongue*—the tongue forms at the same time from this pharyngeal floor and are anatomically associated with thyroid gland by connection through the thyroglossal tract, the lingual remnant of which is known as the foramen caecum.
- *Development of lingual thyroid*—in some cases, thyroid gland fails to descent which gives rise to lingual thyroid. Thus, follicles of thyroid tissue are found in the substance of the tongue.

Clinical Features

- *Age and sex distribution*—females are more affected as compared to males in a ratio of 7:1. The reason for occurrence in female is due to hormonal influence. The age of onset ranges from birth to the 6th decade, with a peak in the 2nd decade.
- *Site*—base of the tongue is the most common site of undescended thyroid gland. It is present in midline.
- *Symptoms*—there may be complaints of dysphagia (difficulty in eating), dysphonia (difficulty in speaking), dyspnea (difficulty in breathing), hemorrhage with pain or feeling of tightness or fullness in the throat. In some cases pain may be referred to ear.
- *Appearance*—it is manifested as a nodular mass in or near the base of the tongue (**Fig. 22-6**), in general vicinity of the foramen caecum. It is often but not always, in the midline.
- *Signs*—the mucosa covering the swelling is thin, smooth and with prominent vascular marking.



Fig. 22-6: Lingual thyroid nodule seen as mass on tongue.

Radiological Features

- *Scintigraphy*—Scintigraphy with ^{131}I and ^{99}Tc can be useful to locate normal and ectopic thyroid tissue. Thyroid radioactive uptake can be determined after administration of ^{131}I .
- *Computed tomography*—it is useful to locate the lingual thyroid. As lingual thyroid nodule contains iodine no contrast agent is used to locate lingual thyroid.
- *Ultrasound*—ultrasound of neck is used to detect lingual thyroid. Doppler ultrasound can also be used. Doppler ultrasound reveals low resistance arterial blood flow.

Diagnosis

- *Clinical diagnosis*—mass seen in vicinity of foramina will give clue to the diagnosis.
- *Radiological diagnosis*—Scintigraphy with iodine will locate the lingual thyroid nodules.

Management

- *Suppressive thyroxin*—suppressive thyroxin should be given for 6 months. This will reduce the size of swelling.
- *Surgical excision*—when it causes difficulty to the patient in spite of thyroxin therapy, excision or ablation with radioiodine is indicated.
- *Surgical excision with reimplantation*—when thyroid gland is absent in the neck, surgical excision with reimplantation can be carried out.
- *Laser*—it can be used as an alternative to scalpel surgery.

Variations in Tongue Movement

- *Curling of tongue*—ability to curl up the lateral borders of the tongue into a tube is noted in 65% of Caucasians and is inherited as an autosomal dominant trait.
- *Folding back tip of tongue*—ability to fold back the tip of the extended tongue, without the aid of the teeth.
- *Trefoil tongue*—clover leaf pattern.
- *Gorlin sign*—extensibility of the tongue, both, forward to touch the tip of nose and backwards into the pharynx.

Tongue Thrusting

Positioning of tongue between the anterior teeth during swallowing, speaking or at rest. It is an infantile swallowing pattern. It may be associated with macroglossia. In these cases, anterior open bite is present.

Patent Thyroglossal Duct Cyst

Development

- *Development of thyroid gland*—thyroid gland develops from an analogue of endothelial cells in the midline of the floor of pharynx, between the first and second branchial arches, just posterior to tubercular impar.
- *Proliferation of cells*—these cells sink into the base of developing tongue descend into the neck and proliferate below the larynx to form thyroid gland.

- *Formation of cyst*—remnants of this part are referred to as thyroglossal duct. Cystic degeneration of it is called as duct cyst.

Clinical Features

- *Age and sex*—it usually occurs in young persons with no sex predilection.
- *Site*—it is seen above the thyroid, in vicinity of the hyoid bone, in midline of the neck.
- *Symptoms*—in some cases, dysphagia may occur.
- *Signs*—it is a firm cystic mass in which formation of fistula may take place. It is compressible.
- *Aspiration*—it yields yellow fluid on aspiration.

Diagnosis

- *Clinical diagnosis*—dysphagia with firm cystic mass in midline of neck will give clue to the diagnosis.
- *Laboratory diagnosis*—the cyst is lined by columnar, respiratory or stratified squamous epithelium. Follicle of thyroid is frequently located in juxtaposition to the cyst lining.

Management

- *Surgical*—it should be surgically excised or enucleated.

Reactive Lymphoid Aggregate

The lingual tonsils, one of the largest oral lymphoid aggregates, are located on the posterior portion of the tongue on the dorsolateral aspect. They are reported to occur on the gingiva, buccal mucosa, tongue and floor of mouth.

Lingual Cyst

It is a rare lesion, located anteriorly in the midline of the tongue (**Fig. 22-7**). It is movable and compressible. It arises as a result of epithelial entrapment during fissural closure of the lateral lingual processes. Surgical excision is recommended.

Local Tongue Disorders

Fissured Tongue

It is also called as '*scrotal tongue*', '*plicated tongue*' and '*lingua dissecta*'.

In this condition tongue with or without a central fissure shows parallel fissures lateral to the midline or fissures at right angle to the long axis of the tongue. A tongue is characterized by furrows, one extending anteroposteriorly and others laterally over the entire anterior surface.



Fig. 22-7: Large lingual cyst of newborn present in the midline of tongue.

Types

- Foliate
- Cribriform
- Plicated.

Etiology

- *Genetic*—fissured tongue is noticed in the generation.
- *Psychological*—it is seen in mentally retarded and psychotic individuals.
- *Trauma*—chronic trauma can lead to fissured tongue.
- *Nutritional deficiency*—in some cases of vitamin deficiency, fissured tongue can occur.

Clinical Features

- *Age*—it can be seen at any age. Number, width and depth of fissures are increased with age in the affected individual.
- *Symptoms*—it is usually painless, except in some occasional cases in which food debris tends to collect in the groove and produce irritation.
- *Appearance*—it is manifested as small furrows or grooves on the dorsal surface, often radiating out from the central groove along the midline of the tongue (**Fig. 22-8**).
- *Pattern*—six different patterns of tongue fissuring were observed, i.e. plication, central longitudinal fissuring (**Fig. 22-9**), double fissures, and transverse fissuring arising from a central fissure, transverse fissuring with a central fissure and lateral longitudinal fissuring (**Fig. 22-10**).
- *Syndrome*—it is associated with Melkersson-Rosenthal syndrome which includes cheilitis granulomatosa, facial paralysis, fissured tongue and non-pitting, non-inflammatory painless edema of face.

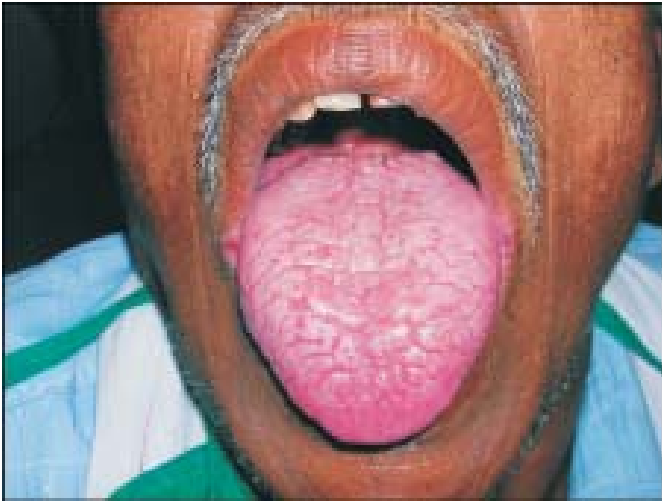


Fig. 22-8: Fissured tongue manifested as furrow radiating from the midline.



Fig. 22-9: Central longitudinal fissure seen in fissure tongue (Courtesy Dr Chole).



Fig. 22-10: Lateral longitudinal fissure seen in fissure tongue (Courtesy Dr Chole).

Diagnosis

- *Clinical diagnosis*—fissure can be easily diagnosed on clinical examination.

Management

- *Maintenance of tongue hygiene*—you have to clean the debris with brush.

Median Rhomboid Glossitis

It is also called as '*central papillary atrophy of tongue*'. It is a developmental defect resulting from an incomplete descent of tuberculum impar and entrapment of a portion between fusing lateral halves of the tongue. It is a benign lesion of the tongue, characterized by rhomboid or oval in shape, changes occur in the tongue mucosa in the midline, just anterior to the foramen caecum (Fig. 22-11).



Fig. 22-11: Rhomboidal shaped lesion seen in tongue (Courtesy Dr Chole).

Pathogenesis

- *Failure of tuberculum impar*—it is a congenital abnormality of the tongue which is due to failure of tuberculum impar to retract or withdraw before fusion of lateral halves of the tongue, so that a structure devoid of papilla is interposed between it.

Etiology

- *Developmental*—the persistent tuberculum impar suggests its developmental origin.
- *Fungal infection*—*Candida albicans* is many times found in this lesion. This type of lesion is called as *posterior midline atrophy candidiasis*.
- *Metabolic*—median rhomboid glossitis is more common in a diabetic than in non-diabetic subjects.

Clinical Features

- *Age and sex*—males predominate over females (3:1). It is common in adults with age ranging from 15 to 84 years.
- *Site*—it is located just anterior to the foramen caecum on dorsal surface of the tongue, in midline and anterior to the circumvallate papillae.
- *Symptoms*—generally asymptomatic, but pain and ulceration are reported. Rest of the tongue may be coated or matted.
- *Appearance*—it appears as an ovoid, diamond or rhomboidal shaped reddish patch or plaque on the dorsal surface of the tongue, immediately anterior to the circumvallate papillae (**Fig. 22-12**). In some cases, flat or slightly raised area stands out from rest of the tongue because it has no filiform papillae.
- *Surface*—the surface is dusky red and completely devoid of filiform papillae and usually smooth. The texture may be varied from a reddish, smooth, granular surface to a lobulated and indurated surface.
- *Kissing lesion*—soft palate erythema may be seen where the lesion of median rhomboidal glossitis touch the palate.



Fig. 22-12: Median rhomboid glossitis
(Courtesy Dr Amit Parate).

Diagnosis

- *Clinical diagnosis*—rhomboidal shaped lesion seen in posterior part of tongue will diagnose the lesion.
- *Laboratory diagnosis*—biopsy shows loss of papillae with varying degrees of hyperparakeratosis. There is proliferation of spinous layer with elongation of rete pegs, which may branch and anastomose.

Management

- *Antifungal agents*—if candidal organism is found, it is treated with an antifungal agent.
- *Long standing cases*—only in long standing cases, cryosurgery or an excisional biopsy is indicated.

Benign Migratory Glossitis

It is also called as 'geographic tongue', 'wandering rash', 'glossitis areata exfoliativa', and 'erythema migrans'.

It refers to irregularly shaped reddish areas of depapillation and thinning of dorsal tongue epithelium which is surrounded by a narrow zone of regenerating papillae that are whiter than the surrounding tongue surface.

Etiology

- *Hypersensitive patient*—in patients with geographic tongue, there is a high frequency of history of asthma, eczema and hay fever.
- *Other factors*—geographic tongue can be seen in immunological reaction, emotional stress, hereditary factors, infections, and nutritional deficiencies.

Classification

- *Type I*—lesion confined to the tongue, with both active and remission phases. No other lesion elsewhere in the oral cavity.
- *Type II*—as type one with similar lesions elsewhere in the mouth.
- *Type III*—lesions on the tongue are not typical and may be accompanied by lesions elsewhere in the mouth. It consists of two forms:
 - *Fixed form*—a few areas of the tongue are affected, but no movement is observed. They may disappear only to recur at the same area.
 - *Abortive forms*—this form starts as yellow-white patches, but disappear before acquiring the typical appearance of geographic tongue.
- *Type IV*—no tongue lesions are present, but geographic areas present elsewhere in the mouth.

Clinical Features

- *Age and sex*—it is common in young and middle aged adults, with an age range of 5 to 84 years with a predilection for females.
- *Site*—lesion confines to dorsal surface (**Fig. 22-13**) and lateral border of the tongue, but may occur on the ventral surface.
- *Size*—it is extremely variable in size and diameter and it may be single or multiple (**Fig. 22-14**).



Fig. 22-13: Geographic tongue seen on dorsal surface of tongue (Courtesy Dr Parate).

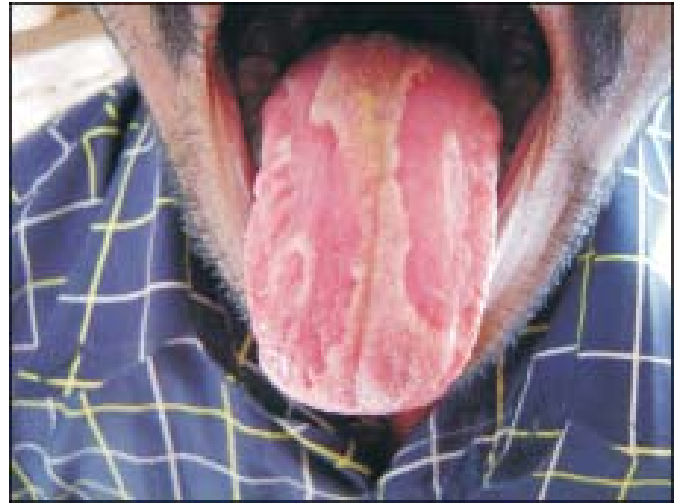


Fig. 22-15: Multiple area of desquamation surrounded by yellowish white line (Courtesy Dr Parate).



Fig. 22-14: Multiple area of erythema seen in geographic tongue.



Fig. 22-16: Geographic tongue elevated fungiform papillae.

- **Symptoms**—it is asymptomatic, but the patient may complain of burning sensation that is made worse by spicy or citrus food.
- **Appearance**—initially appears as a small erythematous, non-indurated, atrophic lesion, bordered by a slightly elevated distinct rim that varies from gray to white to light yellow.
- **Color**—loss of filiform papillae produces pink to red smooth shiny surface except the residual fungiform papillae.
- **Signs**—Multiple areas of desquamation of filiform papillae, in an irregular circinate fashion are seen. Central portion or lesion, sometimes appears inflamed, while the border is outlined by thin yellowish white line or band (**Fig. 22-15**).
- **Fungiform papillae**—fungiform papillae persist in the desquamated areas as small elevated red rods (**Fig. 22-16**).
- **Migration**—area of desquamation remains for a short time in one location and then heals and appears in another location thus giving rise to the term 'migration'.
- **Prognosis**—condition may persist for weeks or months and then regress spontaneously only to recur at a later date.
- **Ectopic geographic tongue**—lesion is not always restricted to tongue and similar irregular or circinate lesions occur elsewhere in oral cavity and are called as *ectopic geographic tongue* or *erythema circinate migrans* or *annulus migrans*.

Diagnosis

- **Clinical diagnosis**—multiple areas of desquamation on dorsal surface of tongue.
- **Laboratory diagnosis**—biopsy shows loss of filiform papillae with hyperparakeratosis and acanthosis. There is also presence of *Monro's abscess*, near the surface.

Differential Diagnosis

- *Psoriasis*—in it, skin lesions are present.
- *Reiter's syndrome*—skin, ocular and urethral lesions occur with arthritis.
- *Pityriasis rosea*—skin lesions present.
- *Lichen planus*—absence of raised whitish yellow rim.
- *Use of strong mouth wash*—history.
- *Anemic condition*—hematological study and absence of raised yellowish white border.

Management

- *For control of burning*—topical local anesthetic agents like lidocaine, dyclonine hydrochloride or diphenhydramine can be given.
- *Other*—bland diet, elimination of irritants and psychological reassurance is useful.
- *Topical therapy*—topical corticosteroids and topical application of salicylic acid and tretinoin (retinoic acid or Vit A) for external use.
- *Zinc supplement*—recent study suggests that zinc supplement is effective in symptomatic geographic tongue.

Hairy Tongue

It is also called as '*lingua villosa, lingua nigra, black hairy tongue*'. It designates an overgrowth of the filiform papillae on the dorsum of the tongue, giving the tongue a superficial resemblance as that of hairiness. There is marked accumulation of keratin on the filiform papillae of the dorsal tongue. There may be delayed shedding of the horny layer of the filiform papillae or there may be an increase in the rate of formation of keratin.

Etiology

- *Fungal and bacterial overgrowth*—overgrowth of fungal organisms like *Candida albicans* and certain bacterial organism can lead to hairy tongue.
- *Use of certain drugs*—oral use of certain drugs (sodium perborate, sodium peroxide and antibiotics like penicillin and Aureomycin).
- *Radiation*—extensive X-ray radiation around head and neck for the treatment of tumors.
- *Poor oral hygiene*—Patients who are unable to maintain oral hygiene can lead to accumulation of keratin in the tongue.
- *After surgery*—it also occurs in patient with intermaxillary fixation and with disturbed orophysiology, due to recent surgery in the oral cavity.
- *Lowered pH*—a lowered pH of oral secretion, which blocks the normal desquamation of epithelial cells covering the filiform papillae.

- *Debilitating diseases*—when the tongue movement is limited by some illness.
- *Foods*—various foods, particularly coffee and tea, probably contribute to the diffuse coloration.

Clinical Features

- *Site*—the lesion involves the dorsum, particularly the middle and posterior one-third.
- *Symptoms*—papillae which are of considerable length will occasionally brush the palate and may produce gagging or bad taste.
- *Signs*—there is hypertrophy of filiform papillae. The papillae may reach a length of 2 cm. Papillae can be elevated by using dental instrument.
- *Appearance*—the elongated papillae have an appearance of hair. The hyperplastic papillae then become pigmented by the colonization of chromogenic bacteria, which can impart a variety of colors ranging from green to brown to black to yellow. This gives it a coated or hairy appearance and retains debris and pigments from substances such as food, tobacco, smoke and candy. Color can be imparted by tobacco, certain food and medicines.
- *Earthy or encrusted tongue*—extreme degree of coating occurring in dehydrated, debilitated and terminally ill patient can lead to a very thick, leathery coating on the tongue which is referred to as 'earthy' or 'encrusted' tongue. It is heavily coated with bacteria and fungi and forms a thickened malted layer.

Diagnosis

- *Clinical diagnosis*—hyperplastic papillae with green to brown color will give clue to the diagnosis.
- *Laboratory diagnosis*—there is elongated papillae with mild hyperkeratosis and inflammatory cells.

Management

- *Maintenance of tongue hygiene*—brushing of the tongue twice daily for 2 minutes, making gentle movements over the involved area towards the tip of the tongue.
- *Elimination of predisposing factors*—all the predisposing factors which are responsible for hairy tongue should be eliminated.
- *Topical keratolytic application*—the topical application of keratolytic agent such as podophyllum in acetone and alcohol suspension seems to be quite effective. It will produce desquamation of hyperkeratotic papillae.

Crenated Tongue

The term is applied to a condition in which indentations of teeth are observed at the lateral margins of the tongue. It

may occur due to abnormal tongue pressure habits and tongue thrusting habits. Any enlargement of the tongue may cause indentations on the teeth.

Often, impression of teeth is seen on the tongue. It is usually an asymptomatic and harmless condition.

Foliate Papillitis

One foliate papilla is present in newborn at each side of the tongue, consisting of 4 to 8 leaves and located at the junction of the anterior two-thirds of the tongue and the base. The folds are separated by grooves of different depths, perpendicular to the longitudinal axis of the tongue. In adults, they are barely noticeable, but sometimes it may swell.

Hypertrophy of lymphoid tissue may be followed by secondary traumatization resulting in so called foliate papillitis.

Clinical Features

- *Age and sex distribution*—it is more common in women, usually in the second half of life.
- *Symptoms*—symptoms may be partly due to upper respiratory tract infection and partly due to irritation. Soreness, tenderness, pain and occasionally, a sour or metallic taste.
- *Location*—the condition may be bilateral with a duration varying from few weeks to many years.
- *Signs*—foliate papillae frequently become inflamed and enlarged, so that it is clinically evident (**Fig. 22-17**).



Fig. 22-17: Foliate papillitis showing inflammation in anterior part of tongue.

Diagnosis

- *Clinical diagnosis*—inflamed and enlarged foliate papillae with pain will aid in diagnosis.

Management

- *Elimination of irritating factors*—it consists of elimination of irritating factors such as sharp edges of teeth and dentures.

Leukokeratosis Nicotina Glossi

It is also called as 'smoker's tongue'. It is homogenous, like leukoplakia with evenly distributed, pinpoint, hemispherical depressions, showing so called golf ball appearance. As a result of heavy smoking, there is loss of papillae. No other clinical features are found in these patients.

Depapillation of the Tongue

Local Causes

- *Eosinophilic granuloma*—it is not related with eosinophilic granuloma of bone. It is also called as 'ulcerated granuloma eosinophilicum diutinum', 'traumatic granuloma' or 'reparative lesion'. The cause of the lesion is unknown. It is characterized by a well demarcated proliferative ulceration covered by thick masses of fibrin and detritus.
 - *Age and sex*—it may occur at any age and does not show a sex predilection.
 - *Site*—it is located on the dorsum, margin or inferior surface of the tongue. Some of the cases are also found on labial mucosa, floor of the mouth, alveolar ridge and gingiva.
 - *Appearance*—the lesions are ulcerative, not indurated and rather well circumscribed.
 - *Symptoms*—there is putrid odor.
 - *Signs*—the ulceration is probably due to moist environment and frequent traumatization. It can be confused with squamous cell carcinoma.
 - *Management*—when one is dealing with an eosinophilic granuloma, spontaneous healing can be expected in a matter of weeks.
- *Traumatic injuries*—the tongue may be repeatedly traumatized, either mechanically or chemically. Trauma is associated with jagged teeth, rough margins of restorations and inadvertent contact of tongue with dental medicaments such as phenol and eugenol. There is localized area of depapillation with papillary regeneration present. A sharp edge of the tooth may cause a yellowish, not indurated and well circumscribed ulcer at the borders of the tongue. Severe damage of the tongue may occur during epileptic seizures. Prolonged oral intubation may cause a large permanent cleft of the tongue. Cotton roll ulcers are rare, but may occur on the borders of the tongue. Such ulcers are not indurated and can be extremely painful.
- *Lesion due to automutilation*—injuries to the tongue can occur due to self inflicted bites. It usually occurs in mentally handicapped persons.
- *Allergic stomatitis*—it refers to edematous changes in part or all of the oral and lingual mucosa, due to

hypersensitivity reaction. It can occur due to certain drugs like antibiotics, cancer chemotherapeutic agent and anticholinergic agents. It can also occur due to variety of allergens such as monomer of the denture, mouthwashes, chewing gum and lipstick. There is edematous swelling of the tongue. There is depapillation of the tongue (**Fig. 22-18**).



Fig. 22-18: Depapillation of tongue is caused due to allergy.

- *Facial hemiatrophy*—it is characterized by unilateral atrophy of the skin, subcutaneous tissues and muscle of the face. There is atrophy of half of the tongue.
- *Cranial arteritis*—rheumatic polymyalgia and temporal arteritis is an inflammatory condition of large and median sized arteries in elderly persons. There is no sex predilection. Symptoms include headache, fever, sweating, malaise, fatigue, anorexia and weight loss. Blindness is the most severe complication. Several cases of painful ulceration and gangrene of the tongue, as a result of arteritis, have been reported. Lingual pain and intermittent blanching of the tongue also has been described. Early use of adequate corticosteroid therapy at the level of 40 to 60 mg daily is required for the treatment of suspected cranial arteritis.
- *Chronic candidiasis*—chronic atrophic candidiasis can be present on the dorsum of the tongue (**Fig. 22-19**). It is difficult to distinguish it from median rhomboidal glossitis. It is diagnosed by scraping and cytological examination.

Systemic Disease

- *Iron deficiency anemia*—inhibition of epithelial reproduction, secondary candidiasis and chronic xerostomia. There are atrophic changes on the dorsum of the tongue (**Fig. 22-20**). It first appears at the tip and lateral borders with loss of filiform papilla. In extreme cases, the entire



Fig. 22-19: Chronic atrophic candidiasis causing depapillation of tongue.



Fig. 22-20: Depapillation of tongue seen in anemia.

dorsum becomes smooth and glazed. The tongue may be very painful and is either pale or fiery red.

- *Plummer Vinson syndrome*—sideropenic anemia shares atrophic glossitis, angular cheilitis, generalized atrophic oral mucosa, oral ulceration and secondary candidiasis. The tongue may be red or pale, painful and fissured. There is also dysphagia and dystrophy of nails.
- *Pernicious anemia*—the patient suffer from general weakness, burning or itching sensation from the oral mucous membrane with disturbance of taste and occasional dryness of mouth. There may be paresthesia, atrophy of filiform and fungiform papillae. In advanced cases, dorsum of the tongue becomes completely atrophic, smooth and fiery red surface. Tongue appears flabby because the normal muscle tonus is reduced.
- *Niacin deficiency*—deficiency of niacin results in a disease called as 'pellagra'. The tongue become fiery red and

devoid of papillae. The filiform papillae are the most sensitive and disappear first. The fungiform papilla may become enlarged. In advanced cases, all the papillae are lost and reddening become intense. In this, tongue may become so swollen that indentation from teeth are found along borders of the tongue.

- *Folic acid deficiency*—there is marked glossitis. The tongue is fiery red and atrophy filiform and fungiform papillae. The tongue is often swollen and small cracks may appear on the dorsum of the tongue.
- *Scleroderma*—fibrosis of submucosal tissue secondary to the obliteration of small vessels by an autoimmune process is responsible for a scarred, shrunken and atrophic appearance of the tongue in scleroderma. In scleroderma the tongue shrinks, losing its mobility and papillary pattern. The color of tongue changes to a vivid appearance due to circulatory disturbances. In the end stages, the tongue lies as a stiff, reduced body in the floor of mouth.
- *Lupus erythematosus*—isolated irregular areas of lingual mucosa, atrophy and ulceration caused by arteritis, are seen in lupus erythematosus.
- *Dermatomyositis*—it is a clinical syndrome consisting of polymyositis associated with skin lesions. The oral mucosa may show dark red or bluish erythema. In the early stages, tongue is markedly swollen and later becomes harder. In the late phase, tongue is atrophic.
- *Diabetes*—decreased nutritional status of the lingual papillae, as a result of vascular changes affecting sub-papillary dorsal capillary plexus supplying it, causes atrophic glossitis. Central papillary atrophy of the dorsum in which low flat papillae are noticed just anterior to the row of circumvallate papillae, is associated with diabetes.
- *Syphilis*—depapillation of the tongue usually occurs in secondary and tertiary syphilis. In secondary syphilis mucus patch occur, which may be single or multiple on the tongue. Tongue in tertiary syphilis may show gumma formation. A more diffuse, chronic, non-ulcerating, irregular induration, with an asymmetrical pattern of grooves and smooth atrophic field covering the entire dorsum is seen. Gumma is often developed in chronic interstitial glossitis. There is atrophy of filiform and fungiform papillae.
- *Zoster infection*—it is a viral infection caused by herpes zoster virus. Numerous vesicles occur on the ventral surface of the tongue.
- *Tuberculosis*—the most frequent involved area is dorsum of the tongue. There is ulceration with irregular outline and undermined borders, covered by yellowish gray fibrinous layer. There is usually pain associated with ulceration.

Neurological Disorders

Glossodynia

Terminology

- *Glossalgia*—the term glossalgia is used to describe painful tongue.
- *Glossopyrosis*—the term glossopyrosis is used to describe burning sensation in the tongue.
- *Lingual paresthesia or dysesthesia*—when just discomfort is felt, it is called as lingual paresthesia or dysesthesia.
- *Stomatodynia*—when the entire oral cavity is involved, the terms stomatodynia, stomatopyrosis and oral dysesthesia are used.

Etiology

- *Local factors*
 - *Habits*—oral habits such as excessive use of tobacco, excessive drinking, frequent uncontrolled movements of the tongue or bruxism.
 - *Dental causes*—local dental causes such as dentures, irritating clasps or a recently fixed bridge.
 - *Referred pain*—referred pain from infected teeth or tonsils, Moeller's glossitis and periodontal disease.
 - *Sensitizers*—it may be caused by peppermint oil in sweets (since it is a volatile oil) and primary and secondary sensitizers.
 - *Local tongue disorders*—geographic tongue, plicated tongue, lichen planus and median rhomboidal glossitis.
 - *Electrogalvanic discharge*—electrogalvanic discharge due to dissimilar metallic restoration.
 - *Atherosclerotic changes*—atherosclerotic changes in the lingual arteries have also been a cause for glossodynia.
 - *Allergy*—allergy to denture base material, metallic restoration and particular food, medication, mouth-washes and dentifrices.
- *Systemic factors*—systemic diseases like multiple myeloma, amyloidosis, muscular tension, hypoestrogenism, pernicious anemia, pellagra, diabetes, vitamin B deficiency can cause glossodynia. Gastric disturbances such as hyperacidity or hypoacidity, angioneurotic edema, mercurialism. Prolonged antibiotic therapy, psychological problems, xerostomia, and hypothyroidism may be responsible for glossodynia.
- *Neurological factors*—trigeminal neuralgia, damage of lingual nerve after surgery, glossopharyngeal neuralgia. Cerebrovascular and subclinical cerebrovascular accidents can possibly cause pain in the tongue.

- *Idiopathic*—when no local, systemic or neurological causes can be detected, the term essential or idiopathic can be used. In these circumstances, depression, cancerophobia and neurosis are regarded as the possible causes.

Clinical Features

- *Nature of pain*—the description varies from patient to patient, some refer it as pain and others as burning, tingling or numbness in the tongue. It may occur as an isolated features or it may be one from group of oral symptoms.
- *Changes in tongue*—it may occur with clinically observable changes in tongue or without observable changes.

Management

- *Removal of local cause*—this is best management treatment for glossodynia.
- *Muscle relaxant*—treatment of muscular problems by correction of malocclusion or by muscle relaxants such as diazepam.
- *Management of systemic problems*—treatment of the systemic cause should be done.
- *Surgical*—surgical exploration of glossodynia with neuropathy.
- *Topical treatment*—use of topical analgesics 0.5% of Diphenhydramine alone or mixed with 0.5% of dyclonine or lidocaine.

Dyskinesia

It is defined as an impairment of voluntary motion, causing movements that are incomplete or only partial.

Generalized Neurological Disease

- *Huntington's chorea*—it is a hereditary disease manifesting progressive degeneration of nerve elements and characterized by choreiform hyperkinesia including grimacing of face. The speech may be affected early, due to the involuntary movements of the tongue.
- *Gilles de la Tourette's syndrome*—it is characterized by motor incoordination and tics, with echolalia and coprolalia. The patient exhibits bizarre facial tics, protrusion of the tongue and the tendency for lip licking.
- *Chorea minor*—it is a disease of the childhood, characterized by the occurrence of rapid irregular, aimless involuntary movements of the muscles of the extremities, face and trunk. The tongue is protruded quickly and rapidly withdrawn to prevent biting of the tongue by involuntary movements of the jaw muscle.

Amyotrophic Lateral Sclerosis

- *Mechanism*—it is a chronic progressive disease of an unknown cause characterized by atrophy and fasciculations of the wasted muscle. It involves demyelination of the corticobulbar tracts and degeneration of cranial nerve motor nuclei.
- *Site*—the muscles of palate, pharynx and tongue are commonly affected.
- *Symptoms*—speech may be slurred. Eating solid food and drinking can cause choking.
- *Early stage*—hypoglossal nerve involvement results in flaccidity, symmetric weakness, slowness of movement and atrophy of tongue.
- *Middle stage*—in the middle stage, a gradual and generalized weakening of the tongue occurs. This is accompanied by spasticity, which results in reduced rate, range and force of articulatory tongue movements.
- *Late stage*—in the late stage, there is virtually unintelligible articulation.

Buccolingual Masticatory Syndrome

- This syndrome consists of repetitive, non-rhythmic abnormal movements, that occur at the speed of normal voluntary movements.
- *Spontaneous dyskinesia*—it consists of movement of mandible and protrusion of the tongue or the lips.
- *Tardive dyskinesia*
 - *Vermicular movements*—fine tremors and fasciculations of the tongue are described as 'vermicular movements'.
 - *Fly-catcher's tongue or bon-bon sign*—rapid darting movements of the tongue is 'fly-catcher's tongue or 'bon-bon' sign.
- *Rabbit syndrome*—when bon-bon sign is associated with smacking movement of the lips with constant lip tremor, it is referred as 'rabbit syndrome'.

Paralysis

It is also called as '*glossoplegia*'. It usually occurs due to unilateral injury of the nucleus in the medulla or the peripheral hypoglossal nerve. There is paralysis and atrophy of the muscles of one-half of the tongue.

Causes

It may be caused by acute anterior poliomyelitis, infectious polyneuritis, neurofibromatosis, syringobulbia, irradiation of head and neck and compression by a tortuous internal carotid artery in the neck.

Clinical Features

- *Signs*—the affected tongue deviates towards the paralyzed side when protruded. The movement towards

the normal side is weak or absent. When the tongue lies on the floor of the mouth, it may deviate or curl slightly toward the healthy side (**Fig. 22-21**) and movements of the tongue towards the back of the mouth on the healthy side, are impaired.

- **Appearance**—if the paralysis is not accompanied by atrophy, the tongue may appear to bulge slightly and to be higher and somewhat more voluminous on the paralyzed side. When atrophy supervenes, paralyzed side becomes smaller and the tongue may become curved towards the paralyzed side with sickle shaped deformities.
- **Location**—in some cases, hypoglossal nerve may be affected bilaterally, causing impairment of the tongue, mobility in lateral direction and atrophy of sides of the tongue.
- **Syndromes associated**—it can be seen in Syndromes like Dejerine (anterior bulbar syndrome), Jackson-MacKenzie (vagoaccessory-hypoglossal syndrome), Collet-Sicard, Duchenne, Möbius and Tapia.



Fig. 22-21: Paralysis of tongue presented as deviation of tongue on the affected side.

Oropharyngeal Dysphagia

It is caused by weakness of the tongue. Symptoms are aspiration while swallowing, regurgitation of food into nose, pharyngeal pain on swallowing and inability of the tongue to move the bolus of food into the pharynx. Dilatation or atony of the pyriform sinuses and pharynx with retention of contrast media in the valleculae is the characteristic radiographic findings.

Dysgeusia and Hypogeusia

Dysgeusia is persistent abnormal taste and hypogeusia is diminished taste sensation. No stimulus is required for the altered tastes, it is called as 'phantom' taste.

Causes

- **Local factors**—local factors like oral candidiasis, oral galvanism, periodontitis, oral lichen planus, and xerostomia.
- **Systemic factors**—systemic factors like vitamin deficiency, nutritional deficiency, food allergy, Sjögren syndrome, liver dysfunction, Crohn's disease, depression, alcoholism, temporal arteritis, migraine, upper respiratory tract infection, chronic gastritis and radiation therapy to neck and head may cause dysgeusia.
- **Pharmacological agents**—pharmacological agents like anticoagulant, antihistamine, antihypertensive, antimicrobial, antipsychotic drugs can cause dysgeusia.

Clinical Features

- **Onset**—onset of dysgeusia is sudden and is distinguished promptly by the patient.
- **Symptoms**—patient may told altered taste as metallic, foul, rancid.
- **Stimulus**—stimulus require for altered taste like certain food and liquids.

Management

- **Removal of underlying cause**—if underlying cause is removed, taste function will return to normal.

Premalignant Lesions and Conditions

Leukoplakia

It can occur anywhere in the oral cavity but tongue is the one of the commonest site. If it occurs on the tongue, it is called as 'chronic superficial glossitis'.

Etiological factors are classically known as 6 'S'. They are: Smoking, Syphilis, Sharp tooth, Sepsis, Sprits and Spices.

Clinical Features

- **Site**—it is confined to the anterior 2/3rd of the tongue (**Fig. 22-23**). It gradually spreads on the dorsum.
- **Appearance**—the surface may become fissure and cracked, due to contraction of the underlying scarred tissue by chronic inflammation.
- **Signs**—the affected area of the tongue shows milky-white patches with cracks and fissures (**Fig. 22-22**). In course of time atrophy tends to succeed hypertrophy, the thickened papillae disappear and the white membrane is worn off. The surface becomes smooth and red.



Fig. 22-22: Milky-white patches seen in leukoplakia of tongue.

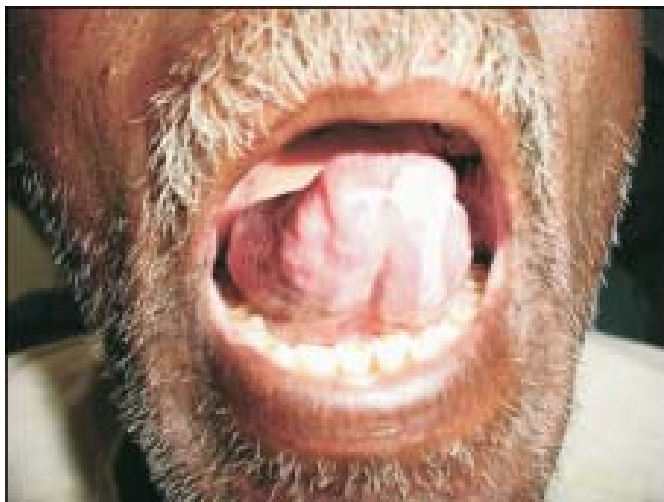


Fig. 22-23: Leukoplakia seen on ventral surface of tongue.

Clinical Staging of Leukoplakia on the Tongue

- *Stage I*—appearance of thin gray transparent film on the affected part of the tongue. The thin milky film may be widespread.
- *Stage II*—the thin film turns opaque and white. In the beginning it looks soft, but later on cracks and fissures appear.
- *Stage III*—hyperplasia causes small nodules and warty outgrowth. Desquamation also appears simultaneously which leaves areas of smooth red and shiny surface.
- *Stage IV*—it is the stage of appearance of clinically detectable carcinoma. The carcinomatous changes usually occur with the fissure. It should be suspected if there is local thickening, bleeding and pain.

Rests of the lesions are described in the Chapter 12: Oral Pre-malignant Lesions and Conditions and Chapter 11: Keratotic and Non-keratotic Lesions.

Malignant Tumor

Squamous Cell Carcinoma

It is the most common oral carcinoma with 60% cases arising from the anterior 2/3rd of the tongue and remainder from the base. Carcinoma of tongue is caused by physical trauma, alcohol, tobacco smoke, candidiasis, syphilis, sepsis, chronic dental trauma and chronic superficial glossitis.

Clinical Features

- *Age and sex*—carcinoma of the tongue is disease of middle and later decades of life, with mean age at presentation being about 60 years. Males are more commonly affected than females.
- *Site*—the majority of tongue carcinoma occurs on lateral border of anterior 2/3rd of the tongue and undersurface of the tongue (Fig. 22-24). The lesions on the posterior border of the tongue are usually of higher grade malignancy, metastasize earlier and often have a poor prognosis. Cancers located in the anterior 2/3rd of the tongue are detected in early stages, as compared to those in the posterior 1/3rd of the tongue.



Fig. 22-24: Carcinoma of tongue occurring on the under surface area.

- *Symptoms*
 - *Painless mass or ulcer*—the most common presenting signs of carcinoma of tongue is a painless mass or ulcer (Fig. 22-25), although in most patients the lesion ultimately becomes painful, especially when it becomes secondarily infected.
 - *Salivation*—excessive salivation gradually appears along with the growth. In late stages, saliva becomes blood stained.
 - *Foetor oris*—as the patient is unable to swallow saliva, offensive smell in the mouth occurs due to bacterial stomatitis.



Fig. 22-25: Ulceration seen on lateral border of tongue in carcinoma.

- *Sore throat*—there is complaint of sore throat and pain in case of lesions on posterior border of the tongue.
- *Immobility of the tongue*—it occurs due to extensive carcinomatous infiltration of the lingual musculature. It becomes worse when floor of mouth is involved and ultimately, it causes difficulty in speech.
- *Hoarseness of voice and dysphagia*—it is present when the carcinoma involves posterior 3rd with involvement of pharynx and larynx.
- *Signs*—carcinoma of the tongue may be seen in four varieties.
 - *Ulcerative variety*—is usually seen near the edge of the tongue. The ulcer looks irregular and the edges are raised and everted. The floor is covered by yellowish gray slough. Base is indurated.
 - *Warty growth*—it usually possesses a broad and indurated base (**Fig. 22-26**). It is developed on excess proliferating growth of filiform papillae. Rarely, it takes cauliflower type of look.



Fig. 22-26: Warty growth seen in carcinoma of tongue (Courtesy Dr Chole).

- *An indurated plaque or mass*—in this case, a typical indurated submucous plaque can be felt (**Figs 22-27A and B**).
- *A fissure*—it is usually presented as a chronic fissure which does not heal.



Figs 22-27A and B: Induration seen in malignancy of tongue (Courtesy Dr Chole).

- *Progress*—the tumor may begin as a superficially indurated ulcer with a slightly raised border and may proceed either to develop a fungating, exophytic mass or to infiltrate the deep layers of the tongue, producing fixation and induration without much surface changes. At an early stage, tongue cancer may appear as thickened, leucoplakic patches, or as a nodule.
- *Complication*—patient usually dies due to cancerous cachexia, starvation, inhalation bronchopneumonia, asphyxias and hemorrhage from involved cervical lymph nodes.

Spread of Carcinoma

- **Local spread**—it spreads by infiltration and invasion.
 - *Carcinoma of anterior 2/3rd of the tongue*—it usually starts on the lateral margins of the tongue and invades the floor of the mouth early, but it does not extend to the other side across the midline.
 - *Carcinoma of posterior 1/3rd of the tongue*—it tends to spread to the corresponding tonsils, epiglottis and soft palate.
- **Lymphatic spread**
 - *Tip of the tongue*—carcinoma of tip of the tongue may drain to the submental nodes, but may also drain to the juguloomohyoid nodes.
 - *Anterior 2/3rd*—it drains into the submandibular lymph nodes.
 - *Posterior 1/3rd*—it drains into jugulodigastric group of the upper deep cervical nodes on both sides of the neck.
- **Blood**—it is rare and extremely late in occurrence. It is only seen when the growth is in extreme posterior part of the tongue.

Management

- **Surgery**—if the growth is less than 1 cm in diameter, it should be removed along with a wide margin of mucosa, not less than 1 cm. If it is localized to anterior 2/3rd of the tongue, partial glossectomy or subtotal glossectomy should be carried out. When the growth reaches within 2 cm of jaw, hemimandibulectomy may be required with excision of the growth.
- **Radiotherapy**—when the growth is more than 1 cm in diameter in anterior 2/3rd, the preliminary treatment is radiotherapy in the form of interstitial radiotherapy.
- **Prognosis**—the 5 years survival rate of cancer tongue is not more than 25%.

Other malignant tumors of the tongue are rare and are discussed in detail in Chapter 16: Malignant Tumor of Jaw.

Pigmentation of Tongue

The tongue may exhibit various patterns of racial melanin pigmentation (**Figs 22-28A and B**).

Endogenous pigmentation is rarely identifiable on the dorsum of tongue because of the thickness of the epithelium, but jaundice may be apparent under the thinner ventral mucosa.

Exogenous pigmentation of filiform papillae of the normal and coated or hairy tongue is very common and result from microbial growth, metabolic products, food debris and dyes from candy, beverages and mouth rinses.

Pigmentation of the tongue has been described by a commonly used anti-chemotherapeutic agents doxorubicin



Figs 22-28A and B: Pigmentation seen in tongue due to melanin deposit.

hydrochloride, which also discolor patient's urine, nailbeds and skin folds. Extravasation of red cells around lingual varicosities may give patchy, bluish red discoloration. The thin tissue overlying a ranula is said to have a greenish blue appearance.

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Classification of Lip Disorders

Developmental

- Congenital lip pits
- Commissural lip pits
- Double lip
- Cleft lip and cleft palate

Cheilitis

- Glandular cheilitis
- Granulomatous cheilitis
- Angular cheilitis
- Contact cheilitis
- Eczematous cheilitis
- Actinic cheilitis
- Exfoliative cheilitis
- Plasma cell cheilitis
- Cheilitis due to drugs

Carcinoma of lip

Miscellaneous

- Chapping of lips
- Actinic elastosis
- Lip ulcers due to caliber persistent artery

Development of Lip

In the sixth week of intrauterine life, two medial nasal processes merge in midline. This will form intermaxillary segment which will give rise to center of upper lip.

In adult, center of upper lip forms *philtrum*. Philtrum is bound laterally by two vertical ridges under the nostril. Lateral part of upper lip fissure presents in maxillary process. This may lead to cleft formation if it is not covered by epithelium and fused. Upper lip is thus formed from one-third medial nasal process and two-third maxillary process.

Anatomy

- *Mucocutaneous junction*—lips are fleshy folds lined by skin externally and mucous membrane internally. The upper and lower lips close along the red margin which represents the *mucocutaneous junction*.
- *Surface*—the lips are covered with skin on the external surface and mucous membrane on the inner surface, which has profuse salivary glands.
- *Extent*—the lip extends from the lower end of the nose to the upper end of the chin.
- *Nasolabial groove*—the upper lip borders onto the nose and is separated from the cheek by a variably deep groove called as *nasolabial groove*.
- *Lower lip*—the lower lip is separated from the chin proper by a more or less sharp and deep groove that is convex superiorly called as *labiomental groove*.
- *Oral commissure*—it is the angle where the upper and lower lip meets.
- *Philtrum*—the upper lip includes the *philtrum*, a midline depression, which can be followed to the nose.
- *Content*—each lip mainly consists of bundles of striated muscle, orbicularis oris, superficial fascia and sub-mucosa. The skin of the lip contains sweat glands, hair and sebaceous gland. The dermal papillae are numerous, with rich capillary supply, which produce reddish pink color of the lips.
- *Vermilion zone*—it is the transitional zone between the skin and the mucous membrane. The vermilion zone contains no hair or sweat glands and contains a few sebaceous glands. In some people, sebaceous glands may be seen as creamy yellow dots. Fordyce's spots along the border between vermilion border and the oral mucosa can also occur.
- *Blood supply of lips*—submental artery to the lower lip and inferior and superior labial arteries to the upper lip.

- *Venous drainage*—anterior facial vein and its branches corresponding to the facial artery provides the venous drainage of lips.
- *Lymphatic drainage*—from central part of lower lip, lymphatics drain to the submental node and rest of the lip to submandibular nodes.
- *Nerve supply*—mental nerve and superior labial nerve.

Developmental Disorders of Lip

Congenital Lip Pits

It is also called as '*paramedian lip pit or congenital fistula*'.

Etiopathogenesis

- *Hereditary*—it is inherited as an autosomal dominant trait.
- *Notching of lip*—there is notching of lips at an early stage of development with fixation of tissues at the base of the notch.
- *Incomplete union*—it may occur due to failure of complete union of embryonic sulci of the lip, resulting in persistent lateral sulci on the embryonic mandibular arch.

Clinical Features

- *Sex predilection*—it is more commonly seen in females.
- *Site*—it is common on vermilion border of either side of midline. It is most commonly seen on lower lip. Lip pits or fistula is unilateral or bilateral depression.
- *Size*—it may be upto 3-4 mm in diameter and may extend as deep as 2 cm.
- *Appearance*—lips, some times appear swollen, accentuating the appearance of the pit.
- *Palpation*—on palpation, sparse mucus secretion may be visible from the base of the lip-pit which occurs due to involvement of underlying minor salivary glands.
- *Syndromes associated*—congenital lip pits may occur in association with *Van der Woude's syndrome* (cleft lip, cleft palate and congenital lip pits). Another syndrome is associated with it is *popliteal pterygium syndrome* which also includes Popliteal webbing (pterygia), cleft lip/cleft palate, genital abnormalities and congenital bands connecting the upper and lower jaws (syngnathia).

Diagnosis

Clinical diagnosis—unilateral or bilateral depression on vermilion border of lip will diagnose these conditions.

Management

- *Surgical excision*—it is done for cosmetic purpose.

Commissural Pits

Commissural lip pits are mucosal invagination occurring at the vermilion border of lip.

Pathogenesis

They occur due to failure of normal fusion of embryonal maxillary and mandibular processes. It is transmitted as autosomal dominant transmission.

Clinical Features

- *Sex distribution*—it is more common amongst males and black people are affected more than white people.
- *Site*—if it is unilateral, it occurs on the right side of the lip.
- *Appearance*—commissural pit appears as a unilateral or bilateral pit at the corner of the mouth on the vermilion surface.
- *Size*—size ranges from a shallow dimple to a tract measuring 4 mm in length and tissue slightly raised above the opening.
- *Palpation*—in squeezing of the lip pit, small amount of saliva can come out.

Differential Diagnosis

- *Congenital lip pit*—it may be associated with facial or palatal cleft.

Management

- *Surgical excision*—it is indicated only in severe condition, where salivary secretion excessive and secondary infection can occur.

Double Lip

It is an anomaly characterized by a fold of excess tissue on the inner mucosal surface of the lip. It may be congenital or acquired because of trauma to the lip.

Pathogenesis

It occurs in 2nd or 3rd week of gestation due to persistent of the sulcus between the pars glabrosa and pars villosa of the lip.

Clinical Features

- *Site*—it usually occurs on inner aspect of upper lip.
- *Cupid bow appearance*—when upper lip is tensed, double lip resembles '*cupid bow*'.
- *Syndromes associated*—it is associated with Ascher's syndrome which consists of double lip, blepharochalasis (it is drooping of the tissue between eyebrow and edge of the upper eyelid so that it hangs loosely over the margin of the lid) and non-toxic thyroid enlargement.

Diagnosis

- *Clinical diagnosis*—cupid bow appearance is typical.

Management

- Surgical excision.

Cleft Lip and Cleft Palate

It occurs along many planes as a result of fault or defect in the development.

Definition

Cleft lip—it is a birth defect that results in a unilateral or bilateral opening in the upper lip between the mouth and the nose. It is also called as harelip. It is wedge shaped defect resulting from failure of two parts of the lip to fuse into a single structure.

Cleft palate—cleft palate is a birth defect characterized by an opening in the roof of the mouth caused by a lack of tissue development.

Development of Cleft

In general, patients with clefts have a deficiency of tissue and not merely a displacement of normal tissue.

- *Unilateral cleft lip*—A cleft lip occurs when an epithelial bridge fails, due to lack of mesodermal delivery and its proliferation from the maxillary and nasal processes. It results from failure of maxillary process on one side to meet and fuse with medial nasal process. This will lead into the division of lip into medial and lateral part. It again occurs due to absence or deficiency of mesodermal masses or their failure to penetrate the ectodermal grooves lead to breakdown of the ectoderm, causing cleft formation. In unilateral cleft lip, the floor of the nose communicates freely with the oral cavity, maxilla on the cleft side is hypoplastic, columella is displaced to the normal side and the nasal ala on the cleft side is laterally, posteriorly and inferiorly displaced.
- *Bilateral cleft lip*—in this, medial mass interposed between two maxillary processes grows down from lateral areas form above the maxillary process. In bilateral cleft lip, the central portion of the alveolar arch is rotated anteriorly and superiorly.
- *Median cleft lip*—it results from partial and complete failure of the medial nasal process to merge. The cleft of lip occurs earlier and inhibits tongue migration, which may then prevent horizontal alignment and fusion of the palatal shelves.
- *Median cleft of mandibular lip*—it occurs due to failure of mesenchymal masses of mandibular process to merge together at 5 weeks of intrauterine life.

- *Cleft of primary palate*—Clefts of the primary palate occur anterior to the incisive foramen. Clefts of the secondary palate are due to lack of fusion of the palatal shelves and always occurs posterior to the incisive foramen. Cleft palate occurs due to disturbances in normal fusion of palatal shelves. This may occur due to failure to unite due to lack of force, interference by the tongue or a disparity in the size of parts involved. It results from failure of the lateral palatine process to meet and fuse with median palatine process. In palatal clefts, the muscles of soft palate are hypoplastic and insert in the posterior margin of the remaining hard palate rather than the midline raphe.
- *Cleft of secondary palate*—The secondary palate closes one week later in females, which may explain why isolated clefts of the secondary palate are more common in females. These are clefts posterior to the incisive foramen. They result of partial or complete failure lateral palatine process to meet, fuse and merge with each other.
- *Cleft of primary and secondary palate*—they result of failure of growth or lack of fusion of three palatine processes with nasal septum and each other.

Etiology

- *Hereditary*—it is one of most important factors to be considered in the etiology.
- *Genetic*—the main possible mode of transmission is by a single mutant gene; producing a large effect or by number of genes (polygenic inheritance), each producing small effects which together create this condition.
- *Nutritional disturbances*—riboflavin deficient diet can produce cleft palate and cleft lip.
- *Developmental*—physiological, emotional and traumatic stress during developmental stages.
- *Defective vascular supply*—defective vascular supply to the area may lead to ischemia which in turn may lead to cleft formation.
- *Mechanical disturbances*—here, the size of tongue may prevent union of the parts.
- *Infection*—infection and lack of inherent developmental force.
- *Miscellaneous*—steroid therapy during pregnancy, alcohol, toxins in the circulation.

Classification

First Classification

- Unilateral incomplete.
- Unilateral complete.
- Bilateral incomplete.
- Bilateral complete.

Second by Veau's

- *Cleft lip*
 - *Class I*—A unilateral notching of vermilion border and it is not extending into the lip.

- *Class II*—a unilateral notching of vermilion with cleft extending into lip but not including the floor of the nose.
- *Class III*—a unilateral cleft of vermilion extending into the floor of the nose.
- *Class IV*—any bilateral cleft of the lip, whether this is complete or incomplete.
- *Cleft palate*
 - *Class I*—involving only soft palate.
 - *Class II*—involving soft and hard palate but not alveolus.
 - *Class III*—involving soft and hard palate and alveolus of one side.
 - *Class IV*—involving both the soft and hard palate and alveolus on both sides of the pre-maxilla.

Third by Kernahan and Stark

- Unilateral incomplete cleft of the primary palate.
- Complete cleft of primary palate ending at the incisive foramen.
- Bilateral complete cleft of the primary palate.
- Incomplete isolated cleft of the secondary palate.
- Complete cleft of the secondary palate—soft and hard palate.
- Unilateral complete cleft of the primary and secondary palate.
- Bilateral complete cleft of the primary and secondary palate.
- Incomplete cleft of the primary palate and incomplete cleft of the secondary palate.

Clinical Features

General

- *Sex*—it is more common in males as compared to females. But incidence of isolated cleft palate is seen more in females.
- *Site*—it is more frequently seen on the left side than on the right side. Left side is involved in 70% of the cases. Cleft of mandibular lip or jaws are rare.
- *Appearance*—a typical patient with cleft palate, cleft lip and ridge exhibits a large defect with a direct opening in the nasal cavity (**Fig. 23-1**).
- *Teeth*—disturbances in the dental structures are seen in this region so that teeth may be missing, deformed, displaced or divided, thus producing supernumerary teeth.

Cleft lip

- There is nasal distortion as lip and nasal tissue pulls towards the attached side.
- *Hare lip*—this term is used to apply for only median cleft lip. Hare lip is derived from the rabbit who normally have cleft in the middle of their lip.



Fig. 23-1: Unilateral cleft lip also involving the cleft palate.

- *Unilateral cleft lip*—a unilateral cleft involves only one side of the lip (**Figs 23-2A and B**).
- *Bilateral cleft lip*—cleft lip is present on the both sides of the midline (**Figs 23-3A and B**).



Figs 23-2A and B: Unilateral cleft lip present on the left side of upper lip.



Figs 23-3A and B: Bilateral cleft lip seen in child patient.

- *Incomplete cleft lip*—it extends for varying distances forward to the nostril, but not upto the nostril (**Fig. 23-4**). The upper part of lip has fused normally.
- *Complete cleft lip*—it extends into nostril and palate is commonly involved. It is often associated with flattening and widening of the nostril of the affected side (**Figs 23-5 and 23-6**).
- *Symptoms*—patient may be presented with difficulty in sucking. Patient also noticed defective speech particularly with the labial letters B, F, M, P and V.
- *Effect on tongue*—there is soft tissue mass between the ends of the bone, uniting the tongue to the lip, so that tongue is bound down.

Cleft palate

- *Site*—there may be cleft of the hard and soft palate or in some cases, cleft of soft palate alone. Entire pre-maxillary



Fig. 23-4: Incomplete cleft lip present on right side which is not extending upto to nostril.



Fig 23-5: Complete cleft lip extending upto nostril.



Fig. 23-6: Complete cleft lip which extends towards the palate.

portion of bone may be missing and in such instances, the cleft appears to be entirely a midline defect.

- *Extent*—cleft of palate may also vary in severity, involving uvula or soft palate (**Fig. 23-7**) or extending all the way through the palate (**Fig. 23-8**) and indirectly to the alveolar ridge on one or both sides (**Fig. 23-9**).



Fig. 23-7: Cleft palate extending all the way towards the palate.



Fig. 23-8: Cleft of palate showing involvement of hard and soft palate.

- *Symptoms*—eating and drinking are difficult due to regurgitation of food and liquid through the nose. Speech problem is serious and tends to increase due to mental trauma.
- *Defect in smelling*—it is due to contamination of the nasal mucous membrane with the oral organism through the cleft palate.
- *Teeth*—the alveolar cleft interferes with the dental lamina. This will result in small upper lateral incisors, absent or even duplicate. Cross bite due to medial collapse of pre-maxilla.



Fig. 23-9: Cleft palate extending to the alveolar ridge

- *Associated anomalies*—isolated cleft palate is associated with other developmental abnormalities like congenital heart disease, polydactyly and syndactyly, hydrocephalus, microcephalus, clubfoot, supernumerary ear, spina bifida, hypertelorism and mental deficiency. Airway problems may arise in children with cleft palates, especially those with concomitant structural or functional anomalies.
- *Syndrome associated*—most common syndrome associated is Pierre-Robin syndrome is the combination of micrognathia, cleft palate and glossoptosis. Affected patients may develop airway distress from their tongue becoming lodged in the palatal defect. Other syndromes which can be present are Goldenhar syndrome, median cleft face syndrome, oral facial digital syndrome, Pert's syndrome, Nagar syndrome, otopalatodigital syndrome, Down's syndrome, and Marfan's syndrome.

Radiographic Features

- It will determine presence and absence of unerupted teeth. The most common missing teeth are maxillary lateral incisors. There is also presence of supernumerary teeth. Teeth are malformed and poorly positioned.
- There is also malposition of the teeth in the region of cleft or in the whole maxilla.
- It will reveal the extent of osseous deformity. There may be complete separation of the premaxilla and maxilla.

Diagnosis

- *Clinical diagnosis*—cleft can be seen clinically on lip and palate.
- *Radiological diagnosis*—cleft palate involving alveolus is seen clearly on radiography.

Management

The complete rehabilitation of the condition requires a multi-disciplinary approach.

- **Cheiloplasty**—it is surgical closure of the lip. A general 'rule of tens' is used in determining optimal timing of lip closure, i.e. 10 weeks of age, 10 pounds of body weight and 10 gm of Hb. At the time of lip closure, when an infant is under general anesthesia, an impression is made for the new obturator.
- **Obturator**—between 3rd and 9th months of age, an obturator is used to provide cross-arch stability, support and to prevent collapse of maxillary arch.
- **Palatoplasty**—it is performed to close an opening in the palate. Surgeons may close the palate in one surgery, when the child is about one year of age or the palate may be closed in two stages, the soft palate is closed first followed by the hard palate.
- **Bone grafting**—sometimes, closure of palatal cleft may be done by bone grafting.
- **Orthodontic therapy**—orthodontic therapy is done to correct malocclusion.
- **Cleft rhinoplasty**—to improve nasal function and correct the distortion.
- **Speech therapy**—speech therapy is given to improve pronunciation of the words.
- **Psychotherapy**—psychological management is necessary.
- **Feeding plate**—to overcome initial feeding problems, feeding plate is used which acts as an obturator to prevent nasal reflux.

Cheilitis

It is inflammation of lip. Various types of cheilitis are described below.

Glandular Cheilitis

It is also called as *cheilitis glandularis*. It is an uncommon condition in which lower lip becomes enlarged, firm and finally everted.

Etiology

- **Sun exposure**—glandular cheilitis can occur due to chronic exposure to sun.
- **Hereditary**—familial occurrence, suggesting a hereditary pattern is also present in glandular cheilitis.
- **Salivary gland inflammation**—inflammation of enlarged heterotopic salivary glands can also be causative factors.
- **Others**—dust, tobacco use and emotional disturbances also been reported in this patients.

Types (Historical)

- **Simple**—multiple, painless, pinhead sized lesions with central depression and dilated canals are present.
- **Superficial suppurative type** (Baelz's disease)—it is characterized by painless swelling, induration, crusting and superficial ulceration of lip.

- **Deep suppurative type** (*cheilitis glandularis apostematosa*, *myxadenitis labialis*)—deep seated infection with abscess and fistula tract that eventually forms a scar.

Clinical Features

- **Age**—it is more common in adults but sometimes, it can also occur in children.
- **Site**—lower lip is involved more often than the upper lip.
- **Symptoms**—enlargement of labial salivary glands occurs which can be nodular.
- **Signs**—orifices of secretory ducts are inflamed and dilated appearing as small red macules over the mucosa.
- **Palpation**—viscid mucous secretion may seep from these openings of everted hypertrophic lips after pressure given on the lip.
- **Volkmann's cheilitis**—it is more severe suppurative form of glandular cheilitis. The lip is considerably and permanently enlarged and is subjected to episodes of pain, tenderness and increased enlargement. The surface is covered by crust (**Fig. 23-10**) and scales beneath which the salivary duct orifice may be discovered.



Fig. 23-10: Severe form of glandular cheilitis showing crust formation

- **Malignant transformation**—it is apparently pre-malignant and epidermoid carcinoma can be associated with it in 18 to 35% of cases.

Diagnosis

- **Clinical diagnosis**—everted hypertrophic lip with secretion after pressure on lip.

Management

- **Vermilionectomy or lip shave**—due to high incidence of associated malignancy, a vermillionectomy or surgical stripping of lips has been recommended. It will give satisfactory cosmetic results.
- **Surgical excision**—if the lips are grossly enlarged, excision of an elongated ellipse of tissue may be required.

Granulomatous Cheilitis or Orofacial Granulomatosis

It is also called as '*Miescher's syndrome*' or '*cheilitis granulomatosa*'. This condition is described in 1945 by Miescher. It is a condition of unknown etiology that is not related to cheilitis glandularis except by the similarity in the clinical appearance of the two diseases.

Etiology

Local causes

- Chronic oral/dental infection
- Embedded foreign material
- Allergy to cosmetics, foods, oral hygiene products and dental restorative materials.

Systemic causes

- Chronic granulomatous disease
- Crohn's disease
- Sarcoidosis
- Tuberculosis

Clinical Features

- *Age and sex*—it is seen in adults as well as in children and there is female predilection.
- *Symptoms*
 - There is diffuse swelling of the lips, especially the lower lip (**Fig. 23-11**).
 - In some cases, an attack is accompanied by fever and mild constitutional symptoms including headache and even visual disturbances.
 - Enlarged lip can create cosmetic problems, difficulty during eating, drinking or speaking.



Fig. 23-11: Diffuse swelling of lower lip in case cheilitis granulomatosis.

- *Signs*—in some cases, scaling, fissuring and vesicles or pustules have been reported.
- *Palpation*—the swelling is usually soft and exhibits no pitting on pressure. Swelling eventually becomes firmer and acquires the consistency of that of hard rubber.
- *Lymph nodes*—the regional lymph nodes are enlarged in some cases, but not always.
- *Skin*—the skin and adjacent mucosa may be of normal color or erythematous.
- *Syndrome*—it is associated with *Melkersson Rosenthal syndrome* which consists of fissured tongue and facial paralysis.

Diagnosis

- *Clinical diagnosis*—soft swelling of lip with fever, headache and vesicle can be seen.

Differential Diagnosis

- *Cheilitis glandularis*—in this condition there is involvement of labial salivary gland.
- *Angioedema*—it is recurrent condition and swelling subsides after giving antihistaminic.
- *Sarcoidosis*—symptoms of fatigue and lethargy are present.
- *Crohn's disease*—gastrointestinal symptoms are present in this case.
- *Lymphangioma*—it is congenital lesion.

Management

- *Corticosteroid injection*—repeated injection of triamcinolone into the lips every few weeks may be effective. Before giving steroids, topical anesthetics gels was applied over the lesion and then 0.1% of triamcinolone acetonide injection is given. This injection should be given weekly for 7 to 10 weeks.
- *Cheiloplasty*—surgical stripping of lip can be done.

Angular Cheilitis

It is also called as '*Perleche*', '*Angular cheilosis*' '*Cheilocandidiasis*'.

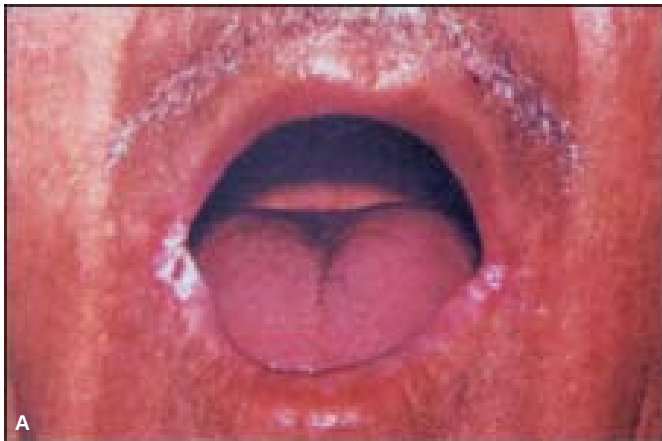
Causes

- *Microorganisms*—particularly candida albicans, but also staphylococci and streptococci.
- *Mechanical factors*—overclosure of jaws such as in edentulous patients or in patients with artificial dentures which lack proper vertical dimensions. In it, folds are produced at the corners of the mouth in which saliva tends to collect and the skin becomes macerated, fissured and secondarily infected. Prognathism may give rise to a similar state of affair in young. The recurrent trauma from dental flossing may occasionally be also implicated.

- *Nutritional deficiency*—it can also occur due to riboflavin, folate and iron deficiency with a superimposed fungal or bacterial infection. General protein deficiency can also cause cheilitis.
- *Diseases of skin*—atopic dermatitis involving the face is often associated with angular cheilitis. The incidence also appears to be increased in seborrhoeic dermatitis.
- *Other factors*—hypersalivation, Down's syndrome, large tongue and constant dribbling being the contributory factors. A rare cause is the presence of a sinus of developmental origin at the angles of the mouth.

Clinical Features

- *Age*—it occurs in young children as well as in adults.
- *Symptoms*—it is characterized by feeling of dryness and a burning sensation at the corners of the mouth.
- *Appearance*—it is usually a roughly triangular area of erythema (**Figs 23-12A and B**) and edema at one or more, commonly both the angles of mouth.
- *Signs*—epithelium at the commissures appears wrinkled and somewhat macerated (**Fig. 23-13**). In time, wrinkling becomes more pronounced to form one or more deep fissures or cracks which appear ulcerated but do not tend to bleed, although a superficial exudative crust may form.



Figs 23-12A and B: Triangular area of erythema seen at the angle of mouth in angular cheilitis.



Fig. 23-13: Macerated lesion seen in angular cheilitis.

- *Rhagades*—linear furrow or fissures radiating from the angle of mouth (rhagades) are seen in more severe forms, especially in denture wearers.
- *Prognosis*—if the lesion is not treated, they often show a tendency for spontaneous remission.

Diagnosis

- *Clinical diagnosis*—triangular area or erythema with wrinkled macerated mucosa at angle of mouth.

Management

- *Removal of the cause*—underlying primary cause should be identified and treated.
- *Nutritional supplement*—a course of vitamin B and iron supplements are useful in these cases.
- *Fusidic acid ointment*—it is used in staphylococcal infection. The lesions should be swabbed first and then fusidic acid ointment or cream should be applied at least four times a day.
- *Miconazole*—miconazole may be preferred, if angular cheilitis is due to candidiasis (cream applied locally together with an oral gel).
- *Gentian violet application*—in some cases, it is useful.

Eczematous Cheilitis

The lips are involved secondary to atopic eczema but possibility of contact dermatitis must also be considered. The management of atopic eczema of the lips is with emollient and topical steroids.

Contact Cheilitis

Definition

Contact cheilitis is an inflammatory reaction of the lips provoked by the irritants or sensitizing action of chemical agents in direct contact with them.

Causes

- **Lipsticks**—they are composed of mineral oils and waxes which form the stick; castor oil as a solvent for the dyes, lanolin as an emollient preservative, perfumes and color. The color includes azo dyes and eosin, which is a bromofluorescein derivative. Sunscreen applied in the form of lipstick can also cause contact cheilitis.
- **Lipsalves and other medicaments**—lipsalves containing lanolin are frequently applied for dryness or chapping. Phenyl salicylates and antibiotics have also been incriminated as a cause of cheilitis.
- **Mouthwashes and dentifrices**—essential oils such as peppermint, cinnamon, clove, spearmint and bactericidal agents can cause cheilitis. Propolis, derived from resin and collected by bees, is a well known sensitizer which has been used in toothpastes.
- **Dental preparations**—mercury and eugenol may cause cheilitis in the absence of stomatitis. Allergy to epimine containing materials used for crowns and bridges can cause cheilitis.
- **Foods**—oranges, mangoes and artichokes are among the food plants which occasionally cause allergic cheilitis and dermatitis of the skin around the lips.
- **Miscellaneous objects**—metal hair clips, metal pencils, cobalt paint on blue pencil can also cause cheilitis.

Clinical Features

- **Site**—lipstick cheilitis is usually confined to the vermillion borders but more often extends beyond that.
- **Signs and symptoms**—there may be persistent irritation and scaling or a more acute reaction with edema and vesiculation (**Fig. 23-14**).

Diagnosis

If acute eczematous changes are obviously present, the diagnosis of contact cheilitis presents no difficulty. If an allergic reaction is suspected, patch test can be carried out.

Management

Topical steroids will give symptomatic relief but the offending substance must be traced and avoided. Most commonly used topical steroids use is 1% triamcinolone acetone.



Fig. 23-14: Contact cheilitis occur due to lipstick showing edema.

Actinic Cheilitis

It is also called as *Actinic cheilosis*. Some other terms which use are *Farmer's lips* or *Sailor's lip* as these people are more exposing to sunlight.

Definition

It is a pre-malignant squamous cell lesion resulting from long-term exposure to solar radiation and may be found at the vermillion border of lip as well as other sun exposed surfaces.

Etiology

- **Chronic sun exposure**—it is the main cause, so it usually occurs in hot, dry regions, in outdoor workers and in fair skinned people.

Clinical Features

- **Site**—the lower lip is more commonly affected than the upper lip as it receives more solar radiation than the upper lip.
- **Age and sex distribution**—it is more commonly seen in adult's patient. It is less common in females due to sunscreen effect of lipstick and less common in blacks due to protective effect of melanin.
- **Signs**—in the early stages, there may be redness and edema but later on, the lips become dry and scaly (**Fig. 23-15**). If scales are removed at this stage, tiny bleeding points are revealed. With the passage of time, these scales become thick and horny with distinct edges. Patient can remove scales but it again reforms within few days. Epithelium becomes palpably thickened with small grayish white plaques. Vertical fissuring and crusting occurs, particularly in the cold weather.
- **Margin**—there is blurring of the margin between vermillion zone and cutaneous portion of lip.



Fig. 23-15: Dryness and scaly nature of patient in actinic cheilitis.

- *Superficial erosion*—at times, vesicle may appear which rupture to form superficial erosions (Fig. 23-16). Secondary infection may occur.
- *Nodule formation*—eventually warty nodules may form which tend to vary in size with fluctuation in the degree of edema and inflammation.



Fig. 23-16: Actinic cheilitis showing ulceration of upper lip in female patient.

- *Signs of malignant transformation*—the possibility of malignancy must always be considered if following features are present;
 - Ulceration in actinic cheilitis.
 - A red and white blotchy appearance with an indistinct vermilion border.
 - Generalized atrophy or focal areas of whitish thickening.
 - Persistent flaking and crusting.
 - Indurations at the base of keratotic lesion.

Diagnosis

- *Clinical diagnosis*—redness, edema with history of chronic sun exposure will give clue to diagnosis.

Management

- *Topical fluorouracil*—for mild cases, application of 5% fluorouracil three times daily for 10 days is suitable. It produces brisk erosion but lips heal within 3 weeks. Application of 5-fluorouracil to the lip will produce erythema, vesiculation, erosion ulceration, necrosis and epithelialization. In some cases, podophyllin is also used.
- *CO₂ snow*—rapid freezing with CO₂ snow or liquid nitrogen on swab stick is used to remove superficial lesions.
- *Vermilionectomy (lip shaves)*—under local anesthesia, the vermilion border is excised by a scalpel and closure is then achieved by advancing the labial mucosa to the skin. Postoperative complications include paresthesia, lip pruritis and labial scar tension.
- *Laser ablation*—carbon dioxide laser therapy has been used to vaporize the vermilion. Good results with no postoperative paresthesia or significant scarring have been reported.
- *Electrodesiccation*—it refers to the deeply penetrating tissue dehydration produced by the insertion of electrodes into the tissue.
- *Prevention*—following management, prevention of recurrence by regular use of sunscreen lip salves is advisable. Liquid or gel waterproof preparation containing para-aminobenzoic acid probably gives the best protection.

Exfoliative Cheilitis

It is also called as *Factitious cheilitis*. It is a chronic superficial inflammatory disorder of the vermilion border of lips characterized by persistent scaling and flaking (Fig. 23-17).



Fig. 23-17: Scaling of lip seen exfoliative cheilitis (Courtesy Dr Shetty).

Causes

- *Chronic injury*—these cases may occur due to repeated lip sucking, chewing or other manipulation of the lips.
- *Personality disorders*—emotional disturbance, psychological difficulties and stress can also lead to exfoliative cheilitis.

Clinical Features

- *Age and sex distribution*—age of occurrence is seen in younger group. Most cases occur in girls.
- *Site*—the process starts in the middle of the lower lip and spreads to involve the whole of the lower lip or both the lips.
- *Symptoms*—the patient complains of irritation or burning and can be observed frequently on biting or sucking the lips.
- *Signs*—it consists of scaling and crusting, more or less confined to the vermillion borders and persisting in varying severity for months or years.
- *Perioral skin*—there is erythema of perioral skin

Diagnosis

- *Clinical diagnosis*—Scaling, crusting with perioral skin erythema will aid to diagnosis.

Management

- *Reassurance and psychotherapy*—this is done to overcome personality disorders. After this, many patients get relief.
- *Topical steroids*—hydrocortisone cream is useful in resolving some chronic cases in some patients.
- *Combination*—hydrocortisone can be combined with iodoquinol (antibacterial and antimycotic) cream can be used in chronic cases of exfoliative cheilitis.
- *Others therapy*—it includes topical silver nitrate, salicylic acid, antibacterial and antifungal formulation.

Plasma Cell Cheilitis

It is an idiopathic benign inflammatory condition characterized by dense plasma cell infiltrate in the mucosa close to the body orifice.

Clinical Features

- *Site*—it can affect penis, vulva, lips, buccal mucosa, palate, gingiva, tongue, epiglottis and larynx.
- *Sign*—it presents as circumscribed patches of erythema (Fig. 23-18), usually on the lower lip in elderly persons.

Diagnosis

- *Clinical*—not possible.
- *Laboratory*—on histopathological examination plasma cell can be seen.



Fig. 23-18: Plasma cell cheilitis presenting as erythematous area on lower lip.

Management

It responds to topical application of powerful steroids or to intradermal injection of triamcinolone.

Drug-induced Cheilitis

Hemorrhagic crusting of the lips is a feature of Steven Johnson syndrome which is commonly caused by drugs but, cheilitis can occur as an isolated feature of a drug reaction- either as a result of allergy or a pharmacological effect.

The aromatic retinoids, etretinate and isotretinoin cause dryness and cracking of lips in most patients.

Carcinoma of Lip

Squamous cell carcinoma is the commonest malignancy to affect the vermillion zone. It occurs in light skinned people who have chronic exposure to sunlight.

Clinical Features

- *Age and sex distribution*—there is peak appearance in 6th and 7th decade of life. It is more common in males as compared to females.
- *Site*—it is most common on the lower lips of fair skinned people and persons who work in outer climate.
- *Onset*—it usually begins on vermillion border of the lip to one side of the midline and it may be covered with crust due to absence of saliva.
- *Actinic cheilitis*—it is preceded by actinic cheilitis which is characterized by innocuous looking white plaque on the lip.
- *Symptoms*—patient may complain of difficulty in speech, difficulty in taking food and inability to close the mouth. There is also pain, bleeding and paresthesia.



Fig. 23-19: Induration and ulceration seen at the angle of mouth (Courtesy Dr Suhas Darvekar).

• **Signs**

- It often commences as a small area of thickening, induration and ulceration or irregularity of the surface (**Fig. 23-19**).
- In some cases, it commences as a small warty growth or fissure on the vermilion border of the lip.
- Crater like lesion having a velvety red base and rolled indurated borders.
- As the lesion enlarged, it takes papillary or an ulcerative form.
- In untreated cases, there is total destruction of lip (**Fig. 23-20**) and invasion of cheek, the gums and the mandible.



Fig. 23-20: Total destruction of lip with extensive growth seen in carcinoma of lower lip.

- **Extent**—papillary lesion grows slowly and infiltrated the deeper tissue relatively late whereas ulcerative growth invade early (**Fig. 23-21**).



Fig. 23-21: Extensive lesion of carcinoma of lip showing destruction of mandible.

- **Metastasis**—it may metastasize and it is usually ipsilateral. Carcinoma of the upper lip metastasizes earlier and more frequently than carcinoma of the lower lip. It involves submaxillary and submental nodes first and then deep cervical nodes. Spread by direct extension into surrounding structures and by metastasis which is through lymphatic channels.

Diagnosis

- **Clinical diagnosis**—ulcerative growth with destruction of lip is present.
- **Laboratory diagnosis**—it is mainly well differentiated malignancies.

Management

- **Surgical**—prognosis is good if the treatment is done before metastasis.
- The best results are seen when being obtained when the entire lip mucosal field is removed for early lesion.

Miscellaneous Disorders

Chapping of the Lips

It is a reaction to adverse environmental conditions in which keratin of the vermilion zone loose its plasticity.

- **Causes**—it is caused by exposure to freezing cold or to hot, dry wind, but acute sunburns can cause very similar changes.
- **Clinical features**—lip becomes sore, cracked and scaly (**Fig. 23-22**). The affected subjects tend to lick the lips or to pick at the scales which make conditions worse.
- **Management**—Management is by application of petroleum jelly and avoidance of the causative environmental conditions.

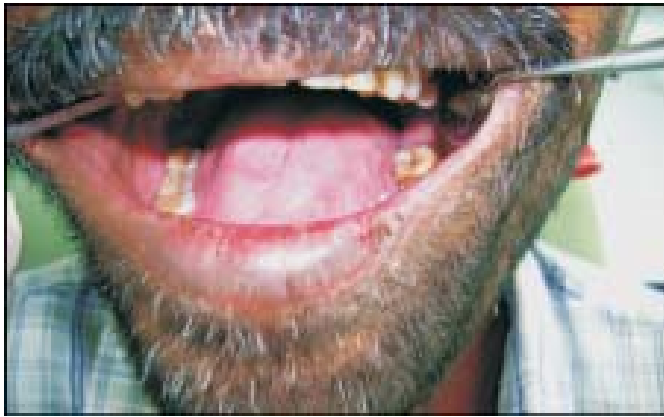


Fig. 23-22: Chapping of lip showing crack in lower lip.

Actinic Elastosis

It is also called as 'Solar elastosis' or 'Senile elastosis'.

Causes

- **Sunlight exposure**—it is caused by prolonged exposure to UV light. UV radiation can produce collagen degeneration in the dermis and extent of this effect is dependent upon factors such as the thickness of stratum corneum, melanin pigment, clothing or chemical sunscreens.

Clinical Features

- **Site**—it is seen on the labial mucosa exposed to sun.
- **Age**—it occurs in elderly population.
- **Signs**—white area of atrophic epithelium develops with underlying scarring of the lamina propria.
- **Appearance**—it includes leathery appearance, laxity with wrinkling and various pigmentary changes.
- **Clinical types**—clinically, it is manifested in three forms:
 - *Cutis rhomboidalis*—thickened skin with furrow giving an appearance of rhomboidal network.
 - *Dubreuilh's elastoma*—diffuse plaque like lesions.
 - *Nodular elastoidosis*—nodular lesion.

Caliber Persistent Artery

A caliber persistent artery is defined as an artery with a diameter larger than normal near a mucosal or external surface. In this condition, main arterial branch extends upto the superficial tissue without reduction in the diameter is present.

Clinical Features

- **Age and sex distribution**—it is more commonly seen in adults as in adults there is loss of tone in the connective tissue.

- **Site**—either lip can involve or some patients have bilateral lesion.
- **Appearance**—the lesion present as linear, arcuate or papular elevation on the lip.
- **Ulcer formation**—such artery in the lip may cause chronic ulceration which can be mistaken for squamous cell carcinoma. The ulcer is attributed to continual pulsation from the large artery running parallel to the surface.
- **Signs**—pulsation can be seen in the lesion. Pulsation is present in lateral direction.

Management

- No treatment is necessary and some time, biopsy is done to avoid the misdiagnosis of the lesion.

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24

Gingival and Periodontal Diseases

Introduction

Gingival and periodontal diseases, in their various forms, have been affecting humans since the dawn of history. Periodontal diseases have been implicated as a major cause of tooth loss besides dental caries.

A review of literature on epidemiology of periodontal disease has concluded that:

- Periodontal disease appears to be a major global public health problem affecting a majority of adult population after the age of 35-40 years.
- The disease starts as gingivitis in young age which, if left untreated, leads to progressive periodontitis.
- More than 90% of the variance of the periodontal severity in the population can be explained by age and oral hygiene.

This concept of periodontitis prevailed until late 1970s. However, the recent studies emphasized on *extent* of the dentition affected by destructive disease (i.e. the percentage of tooth sites involved), and the *severity* of the defects (the amount of lost tissue support). The global distribution of one or several deeper pocket depths (≥ 6 mm) ranged between 1% and 74% in Africa, 8% and 22% in North and South America, 2% and 36% in Eastern Mediterranean, 2% and 40% in Europe, 2% and 64% in South East Asia and 1% and 22% in Western Pacific area.

Normal Periodontium

The periodontium (peri—around, odontos—tooth) consists of gingiva, periodontal ligament, cementum and alveolar bone.

Gingiva

It is the part of the oral mucosa which covers the alveolar process of the jaws and surrounds the neck of the teeth.

The gingiva is divided anatomically into marginal, attached and interdental gingiva.

- *Free gingiva*—the gingiva fits snugly around the teeth, filling each interproximal space between the teeth to the contact area. The gingiva ends in a thin delicate edge called the '*free gingiva*' which is closely adherent to the tooth. The free and attached gingiva blends smoothly with a redder, glossier, unstippled alveolar mucosa of the vestibule and floor of mouth.
- *Color of gingiva*—the color of normal gingiva is a pale coral pink (**Fig. 24-1**) and in the adult the gingiva is dense, firm to touch and insensitive to moderate pressure. It does not bleed easily and it has '*stippled*' or an '*orange peel*' surface.



Fig. 24-1: Normal coral pink color of gingiva.

- *Children gingiva*—in children, gingiva is not stippled and appears redder and more delicate.
- *Interdental tissue*—the interdental tissue in healthy young adults is usually roughly pyramidal in shape, completely filling the space between the teeth almost to the contact area, but there is a small depression or concavity

in the gingival interdental tissue just below the contact area which is called as 'col'. A col lies between the buccal and lingual papillae and is covered with a vestigial structure consisting of epithelial remnants of the enamel organs of the two adjacent teeth.

- **Epithelial attachment**—the most coronal portion of attachment apparatus is called as 'epithelial attachment' or 'epithelial cuff'. It is a band of modified stratified squamous epithelium normally about 0.2 cm in vertical dimension, wrapped around the neck of the erupted teeth in the adults.
- **Fibers of gingiva**—dense fibers of collagen sometimes called as gingival ligament, are seen in gingiva. They are divided into the following groups:
 - **Dentogingival**—it is most numerous groups.
 - **Alveologingival**
 - **Dentoperiosteal**
 - **Circular fibers**—they are small compact group of connective tissue fibers which encircle the teeth. Thus, it helps to maintain the position of the free gingiva.
 - **Transeptal fibers**—they extend from one tooth to other. It is also called as interdental fibers.
- **Blood supply**—it is supplied by vessels of periodontal ligament, supra-periosteal arterioles along the facial and lingual surface of the alveolar bone and arterioles that emerge from the crest of the interdental septa. They are branches of the alveolar arteries and a plexus formed by lingual, buccinator, mental and palatine arteries.
- **Nerve supply**—gingival innervation is derived from fibers arising from nerves in the periodontal ligament and from the labial, buccal and palatal nerve.
- **Venous drainage**—by inferior alveolar vein, posterior superior dental vein and infra-orbital and palatal vein.
- **Lymphatic drainage**—from the upper gingiva passes to the submandibular nodes. Anterior part of the lower gingiva drains into the submental nodes, whereas the posterior part to the submandibular nodes.

Periodontal Ligament

- **Width of periodontal ligament**—the width of periodontal ligament in function is 0.25 to 0.40 mm. Just below the epithelial attachment, there are usually a small number of lymphocytes present.
- **Connective tissue fibers**—a few connective tissue fibers of the periodontal ligament, i.e. the free gingival group fibres project from the cementum of the tooth root into the gingiva without attachment to bone.
- **Composition**—the periodontal ligament is made up of collagen fibers, oxytalan fibers, fibroblasts, amorphous ground substance and interstitial tissue, cementoblasts, osteoblasts and osteoclasts, epithelial rests of Malassez, thin wall blood vessels, lymphatic vessels and tactile sensory nerves.

- **Collagen fibers**—the collagen fibers forming the periodontal ligament are attached to the cementum of the tooth and are inserted into the surrounding tissues. The fibers are arranged in properly defined groups of fibers, also called as principal group. It consists of:
 - **Trans-septal fibers**—it extends interproximally over the alveolar crest and is embedded in the cementum of adjacent teeth.
 - **Alveolar crest fibers**—they are attached coronally, to cementum and at an apical level in bone.
 - **Horizontal fibers**—it runs horizontally from cementum to bone in the same level.
 - **Oblique fibers**—they run from bone, coronally to cementum, apically.
 - **Apical fibers**—from apex of tooth in the bone.
 - **Inter-radicular fibers**—it runs from inter-radicular area of tooth to inter-radicular septa of bone. Seen only in furcated teeth.
- **Frenum attachment**—there are different types of frenum attachment which is described as follows:
 - **Type I**—frenum attached at mucogingival junction.
 - **Type II**—frenum attached in the attached gingiva but not extending till the papilla (**Fig. 24-2**).
 - **Type III**—frenum attached to the papilla, but not extending upto palatal or lingual surface (**Fig. 24-3**).
 - **Type IV**—frenum crosses the papilla and attached to the papilla of lingual or palatal surface (**Fig. 24-4**).

Alveolar Process

It is the portion of maxilla and mandible that forms and supports the tooth sockets. It is formed with eruption of tooth. It provides an osseous attachment to the forming periodontal ligament and disappears after the tooth is lost.

Alveolar process may be defined as the part of the maxilla and mandible that forms and support the socket of the teeth. It is made up of alveolar bone proper, cribriform



Fig. 24-2: Type II frenum attachment.



Fig. 24-3: Type III frenum attachment.



Fig. 24-4: Type IV frenum attachment.

plate and supporting alveolar bone. The latter, in turn, consists of cortical plate which forms the outer and inner plates of alveolar processes and spongy bone which fills the area between these plates and alveolar bone proper.

Alveolar crest is covered with a thin layer of cortical bone and lies within 1 to 1.5 mm of cemento-enamel junction of adjacent teeth. In posterior teeth, alveolar crest is parallel to the cemento-enamel junction. Alveolar crest is continuous with lamina dura with a sharp angle.

Disease of Gingiva and Periodontium

The recent classification for diseases and conditions affecting periodontium was defined by the differences in the clinical manifestations of diseases and conditions. It was the outcome of the consensus opinion arrived at the International Workshop organized by the American Academy of Periodontology in 1999 (Table 24-1).

Table 24-1: Classification of periodontal diseases and conditions

<i>Gingival diseases</i>
<i>Dental plaque-induced gingival diseases*</i>
<i>Non-plaque-induced gingival lesions</i>
<i>Chronic periodontitis**</i>
<i>Localized</i>
<i>Generalized</i>
<i>Aggressive periodontitis**</i>
<i>Localized</i>
<i>Generalized</i>
<i>Periodontitis as a manifestation of systemic diseases</i>
<i>Necrotizing periodontal disease</i>
<i>Necrotizing ulcerative gingivitis (NUG)</i>
<i>Necrotizing ulcerative periodontitis (NUP)</i>
<i>Abscesses of the periodontium</i>
<i>Gingival abscess</i>
<i>Periodontal abscess</i>
<i>Pericoronal abscess</i>
<i>Periodontitis associated with endodontic lesion</i>
<i>Endodontic-periodontal lesion</i>
<i>Periodontal-endodontic lesion</i>
<i>Combined lesion</i>
<i>Development or acquired deformities and conditions</i>
<i>Localized tooth-related factors that predispose to plaque-induced gingival diseases or periodontitis</i>
<i>Mucogingival deformities and conditions around teeth</i>
<i>Mucogingival deformities and conditions on edentulous ridge</i>
<i>Occlusal trauma</i>

* These diseases may occur on a periodontium with no attachment loss or on a periodontium with attachment loss that is stable and not progressing.

** Localized (< 30% of sites involved) and Generalized (> 30% of sites involved). The severity of disease can be characterized on the basis of the amount of clinical attachment loss (CAL) as follows: Slight = 1 or 2 mm CAL; moderate = 3 or 4 mm CAL; and severe ≥ 5 mm CAL and more.

Gingival Diseases

Hereditary Gingival Fibromatosis or Fibromatosis Gingiva

It is also called as 'Elephantiasis gingivae', 'Congenital macrogingivae'. Hereditary gingival fibromatosis is an uncommon condition characterized by diffuse gingival enlargement (Fig. 24-5), sometimes covering major parts or total tooth surfaces. The enlargement develops irrespective of effective plaque removal.

Hereditary gingival fibromatosis may be an isolated entity or part of a syndrome, associated with other clinical manifestations such as hypertrichosis, mental retardation, and epilepsy, hearing loss, growth retardation and abnormalities of extremities. Most cases are related to an autosomal dominant trait. The most common syndrome of hereditary gingival fibromatosis includes hypertrichosis, epilepsy and mental retardation.



Fig. 24-5: Fibromatosis gingivae showing diffuse overgrowth (Courtesy Dr Parate).

Clinical Features

- **Age**—it may be present at birth or may become apparent with eruption of deciduous or permanent teeth.
- **Appearance**—it is manifested as dense smooth diffuse or nodular overgrowth of gingival tissue of one or both arches that usually occurs at the time of eruption of teeth (**Figs 24-6A and B**). It has a characteristic pebbled surface. In some cases, surface has a nodular appearance.
- **Color**—tissue is of normal or pale color. In some cases, it may appear pink.
- **Consistency**—it is often so firm, leathery and dense that it prevents normal eruption of teeth.
- **Symptoms**—it is not painful and shows no tendency for hemorrhage.
- **Extent**—extent may be such that the crown of fully erupted teeth may be hidden.
- **Significance**—the dense firm gingival swelling results in varying spacing between the teeth and change in profile and facial appearance.
- **Radiological features**—in the radiograph, significant bone loss can be present. At the same time, soft tissue is also visible (**Fig. 24-7**).

Diagnosis

- **Clinical diagnosis**—diffuse fibrous overgrowth is seen clinically and it is readily diagnosed.
- **Laboratory diagnosis**—epithelium is thickened with elongation of rete pegs, although the bulk of the tissue is composed of dense fibrous connective tissue. Bundles of collagen fibers are coarse and show few interspersed fibroblasts or blood vessels.



Figs 24-6A and B: Fibromatosis gingivae preventing eruption of teeth in the oral cavity.



Fig. 24-7: OPG of patient having gingival fibromatosis.

Management

- *Surgical*—surgical removal of excessive tissue with exposure of teeth is necessary. However, recurrences are common.
- *Extraction of teeth*—dramatic improvements in gingival enlargements have been observed following extraction of teeth.

Gingivitis

It is the inflammation of the gingiva.

Pathogenesis of Gingivitis (plaque associated)

There is increase in numbers of normal flora organisms that (*Streptococcus*, *Capnocytophaga*, *Actinomyces*, etc.) grow in the gingival crevice producing toxins that cause an inflammatory reaction in the gums. Plaque accumulates in the crevice resulting in an inflammatory reaction. Complications include development of periodontitis.

Classification

Gingivitis can be classified on the basis of:

- *Course and duration*:
 - *Acute gingivitis*—it is of sudden onset and of short duration and can be painful. A less severe form of acute gingivitis has been termed subacute.
 - *Recurrent gingivitis*—recurrent gingivitis reappears after having been eliminated by treatment or disappearing spontaneously.
 - *Chronic gingivitis*—chronic gingivitis is slow in onset and is of long duration. It is painless unless complicated by acute or subacute exacerbations.
- *Distribution*:
 - *Localized gingivitis*—it is confined to the gingiva of a single tooth or a group of teeth (**Fig. 24-8**).
 - *Generalized gingivitis*—generalized gingivitis involves the entire mouth (**Fig. 24-9**).
 - *Marginal gingivitis*—marginal gingivitis involves the gingival margin and may also involve a portion of the contiguous attached gingiva.
 - *Papillary gingivitis*—papillary gingivitis involves the interdental papilla and often extends into the adjacent portion of the marginal gingiva (**Fig. 24-10**).
 - *Diffuse gingivitis*—diffuse gingivitis involves the gingival margin, the interdental papilla and the attached gingiva.

Stages of Gingivitis

- *Stage I gingivitis* (the initial lesion)—the first manifestation of gingival inflammation is vascular changes, consisting essentially of dilation of capillaries and



Fig. 24-8: Localized gingivitis involving lower anterior region.



Fig. 24-9: Generalized gingivitis showing involvement of entire gingiva.



Fig. 24-10: Papillary gingivitis involving interdental papilla.

increased blood flow. No clinical signs appear in this stage. It occurs within 2-4 days of plaque accumulation.

- *Stage II gingivitis* (the early lesion)—this stage is usually seen following 4-7 days of plaque accumulation. In this stage, clinical signs of erythema appear, mainly owing to the proliferation of capillaries and increased formation of capillary loops between rete pegs or ridges. Bleeding on probing may also be evident.
- *Stage III* (the established lesion) (**Fig. 24-11**)—the lesion develops generally after 14-21 days of plaque accumulation. Blood vessels become engorged (**Fig. 24-12**) and congested, venous return is impaired and blood flow becomes sluggish. The established lesion can be described as moderately to severely inflamed.



Fig. 24-11: Established gingivitis is moderately inflamed.



Fig. 24-12: Stage III gingivitis showing engorgement of blood vessels.

- *Stage IV gingivitis* (the advanced lesion)—the lesion extends into the alveolar bone and known as the phase of periodontal destruction.

Clinical Features of Gingivitis

- *Symptoms*—the earliest symptoms of gingival inflammation are increased gingival fluid production and bleeding from the gingival sulcus on gentle probing.
- *Color*—gingiva becomes redder when there is an increase in vascularization or when the degree of epithelial keratinization is reduced. The color becomes paler when vascularity is reduced and keratinization is increased. The color may be sometimes reddish blue or deep blue, if venous stasis has occurred. The color change starts in the interdental papillae, gingival margins and spreads to the attached gingiva.
- *Consistency*—the consistency of gingiva may be spongy that pits on pressure and there may be marked softness of the gingiva.
- *Enlargement of gingiva*—sometimes in inflammation, there may be gingival enlargement and it may lead to change in the contour of gingiva.

Plaque induced gingivitis without local contributing factors

Characteristic of plaque-induced gingivitis are discussed in Table 24-2.

Table 24-2: Characteristics common to all plaque-induced gingival diseases

- Signs and symptoms are confined to gingiva.
- The presence of dental plaque to initiate or exacerbate the severity of the lesion.
- Clinical signs of inflammation (enlarged gingival contours due to edema or fibrosis, color transition to a red or blue-red hue, elevated sulcular temperature, bleeding upon provocation, increased gingival exudates).
- Clinical signs and symptoms associated with stable attachment levels on a periodontium with no loss of attachment or on a stable but reduced periodontium.
- Reversibility of the disease by removing the etiology(ies).
- Possible role as a precursor of attachment loss around teeth.

Plaque-induced gingivitis with local contributing factors

- *Teeth anomalies*—tooth anomalies such as enamel pearls and cemental tears can modify or predispose to plaque-induced gingival diseases. Enamel pearls are found in 1.1-5.7% of molar teeth and maxillary second molar are most commonly involved.
- *Dental restoration*—subgingival margin of restorations and violation of biologic width can affect the health of adjacent gingival tissue.
- *Root fracture*—root fracture and root perforations during endodontic therapy are associated with inflammation of the gingiva and sometimes, pocket formation due to enhanced plaque accumulation.
- *Cervical root resorption*—cervical root resorption may result in inflammation particularly when a communication is established with the sulcus.

- *Other causes*—other cause like deep bite (**Fig. 24-13**), palatogingival groove (**Fig. 24-14**) and shallow vestibule (**Fig. 24-15**) can also lead to gingival inflammation.



Fig. 24-13: Deep bite causing gingival disease.



Fig. 24-14: Palatogingival groove causing gingival disease.



Fig. 24-15: Shallow vestibule causing gingival disease.

Gingivitis due to systemic factors

- *Puberty associated gingivitis*—it appears to be exaggerated response of gingival tissues towards high levels of hormones, especially estrogens and testosterone, secondary to plaque accumulation.
- *Menstrual cycle associated gingivitis*—although bright red hemorrhagic lesions have been described prior to the onset of menses, clinically detectable changes do not seem to be associated with the menstrual cycle. However, an increase in gingival fluid by 20% has been described in 75% of women during ovulation.
- *Pregnancy-associated gingivitis*—the exaggerated response of gingiva to local factors can often appear during the second trimester and regress upon parturition. The condition can be reversed with suitably high levels of plaque control. Pyogenic granuloma of pregnancy is a localized mass of highly vascularized tissue arising as an exaggerated response to plaque. It commonly arises from the proximal gingival tissues and has a pedunculated base. It may show ulceration of its thin epithelial lining. Bleeding on mastication or spontaneously may be a presenting complaint.
- *Diabetes mellitus-associated gingivitis*—there is evidence that Type I diabetes in children is associated with gingival changes. However, when this type of diabetes is under good control, the gingival inflammation is less exaggerated.
- *Leukemia-associated gingivitis*—the leukemia has been associated with gingival changes including gingival swelling, redness/blueness, sponginess and a glazed appearance. The leukemia most commonly associated with gingival changes is acute myeloid leukemia. Any other condition affecting the platelet deficiency can affect periodontium. Cyclic neutropenia, a condition characterized by fluctuation in neutrophils count with a periodicity of 14-36 days, is associated with oral ulceration and exaggerated gingival response to dental plaque.
- *Drug influence gingivitis*—the increased use of oral contraceptives by postmenopausal women has been associated with a higher incidence of gingival inflammation and development of gingival enlargement. This phenomenon can be reversed by discontinuation of medications.
- *Ascorbic acid deficiency gingivitis*—severe vitamin C deficiency in humans results in scurvy, a disease characterized by hemorrhagic diathesis and retardation of wound healing. The clinical features include bleeding, swollen gums and loosened teeth. Scurvy results in defective formation and maintenance of collagen, retardation or cessation of osteoid formation and impaired osteoblastic function. It is also characterized by increased capillary permeability, susceptibility to

traumatic hemorrhages, hyporeactivity of the contractile elements of the peripheral blood vessels and sluggishness of blood flow.

Management of Gingivitis

- *Removal of local irritant*—the local irritants should be removed at this stage.
- *Plaque control*—thorough plaque control should be done with scaling and polishing.
- *Chlorhexidine mouth wash*—use of chlorhexidine, on a short-term basis.

Plasma Cell Gingivitis

It is also called as 'Atypical gingivostomatitis' or 'Atypical gingivitis'. It is allergic in origin and is caused by some ingredients in chewing gums, dentifrices or various diet components.

Clinical Features

- *Sex*—it is more frequently in women and young adults.
- *Site*—it is located on the oral aspect of the attached gingiva and therefore, differs from plaque induced gingivitis.
- *Symptoms*—patients may complain of a sore and burning mouth, scaling of lips and angular cheilitis.
- *Appearance*—entire free and attached gingiva is edematous, friable, granular, bright red (**Fig. 24-16**) and bleeds on slightest provocation.



Fig. 24-16: Bright red color inflammation seen in plasma cell gingivitis.

Diagnosis

- *Clinical diagnosis*—bright red color inflammation with history of some allergen.
- *Laboratory diagnosis*—lamina propria is infiltrated by plasma cells.

Management

- *Cessation of exposure*—cessation of exposure to allergen resolves the lesion.

Gingival Enlargement

Classification

- *Inflammatory enlargement*—it can be acute or chronic.
- *Drug induced*—may occur due to dilantin, nifedipine or cyclosporine.
- *Combined*—it is combination of (inflammatory + fibrotic).
- *Enlargement due to systemic disease*—many systemic diseases like leukemia, Crohn's disease can cause gingival enlargement.
- *Neoplastic enlargement*—benign and malignant tumor of gingiva can cause enlargement.

Acute Inflammatory Enlargement or Gingival Abscess

It results from bacteria carried deep into the tissue, when a foreign substance such as a toothbrush bristle, a piece of apple core or a lobster shell fragment is forcefully embedded into the gingiva.

Clinical Features

- *Site*—it is limited to the marginal gingiva or the interdental papilla (**Fig. 24-17**).
- *Onset*—it is localized, painful, rapidly expanding lesion that is usually of sudden onset.
- *Appearance*—in early stages, it appears as a red swelling with a smooth, shiny surface. Within 24 to 48 hours, the lesion usually becomes fluctuant and pointed with a surface orifice, from which an exudate may be expressed.
- *Teeth*—the adjacent teeth are sensitive to percussion.



Fig. 24-17: Acute gingival inflammatory enlargement seen in lower anterior region.

Diagnosis

- *Clinical diagnosis*—red swelling with shiny surface associated with pain will give clue to the diagnosis.
- *Laboratory diagnosis*—it consists of a purulent focus in the connective tissue, surrounded by a diffuse infiltration of polymorphonuclear leukocytes, edematous tissue and vascular engorgement.

Management

- *Incision*—after topical anesthetic is applied, the fluctuant area of the lesion is incised with a blade and the incision is gently widened to permit drainage. The area is cleansed with warm water and covered with a gauze pad. After the bleeding stops, patient is instructed to rinse every 2 hours with a glassful of water.

Chronic Inflammatory Enlargement

Etiology

- *Local factors*
 - It can be caused by prolonged exposure to dental plaque, which may occur due to poor oral hygiene (**Fig. 24-18**), abnormal relationship of adjacent and opposing teeth, lack of tooth function, overhanging margins of dental restoration and improperly contoured dental restoration or pontics.
 - Food impaction, irritation from clasps or saddle areas of removable prosthesis and nasal obstruction.
- *Habits*—such as mouth breathing can cause gingival enlargement, which is more common in the anterior region.



Fig. 24-18: Inflammatory gingival enlargement occurs due to calculus and poor oral hygiene.

Clinical Features

- *Site*—the enlargement is generally papillary or marginal and localized or generalized.

- *Appearance*—it originates as a slight ballooning of the interdental papilla or the marginal gingiva. In early stages, it produces a life preserver-like bulge around the involved teeth. It increases in size, until it covers a part of the crown.
- *Color*—lesions may be deep red (**Fig. 24-19**) or bluish red. They are soft and friable, with a smooth shiny surface and tendency to bleed.



Fig. 24-19: Inflammatory enlargement shows deep red in color.

- *Progress*—it progresses slowly and painlessly, unless complicated by acute infection or trauma.
- *Discrete mass*—sometimes, enlargement can occur as a discrete, sessile or pedunculated mass, resembling a tumor (**Fig. 24-20**). They may undergo spontaneous reduction in size, followed by exacerbation and continued enlargement. Painful ulceration, sometimes, occurs between the mass and the adjacent gingiva.



Fig. 24-20: Inflammatory gingival enlargement presented as discrete mass.

Diagnosis

- *Clinical diagnosis*—inflammatory enlargement with ballooning of papilla around the teeth will give clue to diagnosis.
- *Laboratory diagnosis*—there is proliferative features of chronic inflammation. Lesions may contain preponderance of inflammatory cells with vascular engorgement.

Management

- *Removal of local factors*—all the factors which are responsible for enlargement should be removed.
- *Antibiotics*—antibiotics should be given. The most commonly used antibiotics is metronidazole.
- *Flap surgery*—periodontal flap surgery is done if necessary.

Drug Influence Gingival Enlargements

Dilantin Sodium

Dilantin sodium is an anticonvulsant drug, which is used to control of epileptic seizures.

Clinical Features

- *Onset*—gingival hyperplasia may begin as early as two weeks after dilantin therapy.
- *Site*—the hyperplasia is generalized throughout the mouth, but it is most severe in maxillary and mandibular anterior region.
- *Appearance*—the first change noted is a painless bead like enlargement of the gingiva, starting with one or two interdental papillae (**Figs 24-21A and B**). The surface of gingiva shows an increase in stippling and finally, a cauliflower, warty or pebbled surface. As the enlargement increases, the gingival tissue becomes lobulated and clefts are seen between each enlarged gingiva.
- *Palpation*—palpation reveals that the tissue is dense, resilient and insensitive. It shows little tendency to bleed.
- *Significance*—they may develop massively, covering a considerable portion of the crown. They may interfere with occlusion. The presence of an enlargement makes plaque control difficult, resulting in a secondary inflammatory process that complicates the gingival hyperplasia. In hyperplasia associated with Dilantin sodium the presence of dental plaque does not seem to initiate the enlargement. Additional studies have demonstrated that oral hygiene procedures may limit the severity of the lesion but are unable alone to lead to a reversal of the condition.



Figs 24-21A and B: Dilantin-induced gingival enlargement showing bead-like enlargement.

Cyclosporine

It is a potent immunosuppressive agent used to prevent organ transplant rejection and to treat several diseases of autoimmune origin.

Clinical Features

- *Clinically*, it is similar to that induced by Dilantin.
- *Site*—growth starts in the interproximal papillae, more frequently in anterior facial areas, partially covering the crown.
- *Appearance*—the tissue is usually pink, dense and resilient, with stippled or granular surface and little bleeding tendency (**Fig. 24-22**).

Nifedipine

- *Action*—it is a calcium channel blocker that induces direct dilatation of the coronary arteries and arterioles, improving the oxygen supply to the heart muscles. It is used in the treatment of acute and chronic coronary insufficiency.



Fig. 24-22: Drug-induced gingival enlargement showing pink resilient surface due to cyclosporine administration.

- Gingival overgrowth occurs in 20% of the cases. The clinical and histological features are same as seen in Dilantin hyperplasia.

Diagnosis

- *Clinical diagnosis*—enlargement of the gingiva with history of drugs will diagnose the condition (Fig. 24-23).
- *Laboratory diagnosis*—the stratified squamous epithelium covering the tissue is thick and has a thin keratinized layer. The rete pegs are extremely long and thin, sometimes called as 'test tube' pegs, with considerable confluence. Mitotic figures are seldom seen.



Fig. 24-23: Nifedipine-induced gingival enlargement.

Management

- *Surgical excision*—If hyperplasia interferes with function, surgical excision is recommended.
- *Discontinue the drug*—discontinuing the drug will result in gradual diminution of the bulk of the gingiva.

Combined Gingival Enlargement

In this type, enlargement of both inflammatory as well as fibrous component are present. It can be localized (Fig. 24-24) or generalized (Fig. 24-25).



Fig. 24-24: Localized combined gingival enlargement showing inflammatory and fibrous component.



Fig. 24-25: Generalized combined gingival enlargement.

Pregnancy-induced Gingival Enlargement

It can be marginal or generalized and may occur as a single or multiple tumors like masses.

Clinical Features

- *Marginal enlargement*—it tends to be more prominent interproximally, than on the facial and lingual surfaces. The enlarged gingiva is bright red or magenta, soft and friable and has a smooth shiny surface. Bleeding occurs spontaneously or on slight provocation.
- *Pregnancy tumor*—it is an inflammatory reaction to the local irritants. It usually appears after 3rd month of pregnancy.

- **Appearance**—the lesions appear as discrete, mushroom like (**Fig. 24-26**), flattened spherical masses, that protrude from the gingival margins or more frequently from the interproximal space and are attached by sessile or pedunculated base. It tends to expand laterally. The pressure from the tongue and the cheek perpetuates its flattened appearance.
- **Color**—it is generally dusky red or magenta; it has a smooth glistening surface that frequently exhibits numerous deep red, pinpoint markings.
- **Consistency**—the consistency varies from semifirm, but may have a varying degree of softness and friability.
- **Symptoms**—it is usually painless, unless its size and shape foster the accumulation of debris under its margin or interfere with occlusion, in which painful ulceration may occur.



Fig. 24-26: Mushroom like enlargement seen in pregnant women.

Diagnosis

- **Clinical diagnosis**—mushroom like enlargement in pregnancy will diagnose this condition.
- **Laboratory diagnosis**—the connective tissue consists of numerous, diffusely arranged, newly formed and engorged capillaries, lined by cuboidal endothelial cells. The epithelium exhibits some degree of intracellular or extracellular edema with prominent intercellular bridges and leukocytic infiltration.

Management

- **Maintenance of oral hygiene**—most gingival enlargement during pregnancy can be prevented by the removal of local irritants and institution of a fastidious oral hygiene.

Puberty-induced Gingival Enlargement

It is usually associated with inflammatory type.

Clinical Features

- **Age and sex**—it occurs in both, males and females in pubertal age group. It also appears in areas of local irritants.
- **Site**—it involves mainly the marginal gingiva and interdental gingiva.
- **Appearance**—it is characterized by prominent bulbous interproximal papillae (**Fig. 24-27**). Sometimes, only facial gingivae are enlarged, as the mechanical action of the tongue prevents heavy accumulation of local irritants on the lingual surface.



Fig. 24-27: Puberty-induced gingival enlargement showing prominent bulbous papillae.

Diagnosis

- **Clinical diagnosis**—bulbous enlargement at the onset of puberty will give clue to the diagnosis.
- **Laboratory diagnosis**—microscopic picture is that of chronic inflammatory cells with prominent edema and associated degenerative changes.

Management

- **Removal of local irritant**—after puberty, the enlargement undergoes spontaneous reduction, but does not disappear until local irritants are removed.

Necrotizing Ulcerative Gingivitis

It is an endogenous oral infection that is characterized by necrosis of gingiva. It is also called as 'Trench mouth' due to its prevalence in combat trenches (**Fig. 24-28**). Other synonyms for this are 'Vincent's infection', 'Acute ulceromembranous gingivitis', 'Fusospirochetal gingivitis' and 'Acute ulcerative gingivitis'. Tissue destruction is caused by endogenous organisms that act either on the tissue or, indirectly by triggering an inflammatory reaction.

Etiology

- *Role of bacteria*—it is caused by fusiform bacilli and spirochetes. In addition to it, bacteroides intermedius is also responsible for ANUG.
- *Local predisposing factors*—poor oral hygiene, pre-existing marginal gingivitis and faulty dental restorations. Deep periodontal pockets offered favorable environment for occurrence of the disease. Area of gingiva is traumatized by opposing maloccluded teeth. As tobacco smoke has a direct toxic effect on the gingiva, smoking and emotional stress can predispose for ANUG.
- *Systemic predisposing factors*
 - *Nutritional deficiency*—nutritional deficiency like vitamin C, vitamin B₂ accentuate the severity of the pathologic changes induced by the fusospirochetal bacterial complex.
 - *Debilitating disease*—chronic diseases like leukemia, aplastic anemia, syphilis, severe gastrointestinal disturbances and AIDS can act as predisposing factors.
 - *Marked malnutrition*—it may be predisposing factors to necrotizing ulcerative gingivitis.
 - *Psychosomatic factors*—the disease often occurs in association with a stress situation as well as with increase in adrenocortical secretion.

Clinical Features

- *Age*—it is most commonly seen in the age group of 16 to 30 years, but can be seen in children from a low socioeconomic group, in underdeveloped countries.
- *Symptoms*—onset is sudden with pain, tenderness, profuse salivation and peculiar metallic taste. Spontaneous bleeding from gingival tissue occurs. There is also a loss of sense of taste and diminished pleasure from smoking. The typical fetid odor ultimately develops, which may be extremely unpleasant.
- *Signs*—teeth seem slightly to be extruded and are sensitive to pressure or have a woody sensation. They are slightly movable and the patient is unable to eat properly. Gingiva may become superficially stained with brown color. There is blunting of interdental papillae (Fig. 24-28).
- *Appearance*—a typical lesion consists of necrotic punched out, crater like ulcerations are developed most commonly on the interdental papillae and marginal gingiva (Fig 24-29). Removal of the lesion leaves raw surface. The surface of gingival crater is covered by a gray, pseudo-membranous slough, demarcated from the remainder of the gingival mucosa by pronounced linear erythema. In some cases, ulceration may develop on cheek, lip, tongue, palate and pharyngeal area. If untreated, it may result in progressive destruction of the periodontium and



Fig. 24-28: Necrotizing ulcerative gingivitis showing blunting of interdental papilla.

denudation of the roots, accompanied by increase in the severity of complications.

- *Lymph nodes*—regional lymph nodes are enlarged.
- *Fever*—there may be a slight elevation of temperature.
- *Systemic complications*—in severe cases, there may be systemic complications like high fever, increased pulse rate, loss of appetite and generalized lassitude.



Fig. 24-29: Necrotizing ulcerative gingivitis showing ulceration of marginal gingiva.

Clinical Classification of NUG

- *Stage I*—only the superior margins of the interdental papillae are affected. There is marked tendency of bleeding on probing.
- *Stage II*—the process spreads to the marginal gingiva with characteristic punched out destruction.
- *Stage III*—the attached gingiva is affected in addition to the interdental papillae and marginal gingiva.
- *Stage IV*—the necrotizing process has resulted in denudation of bone.

Diagnosis

- *Clinical diagnosis*—punched out ulceration of gingiva with systemic features will give clue to the diagnosis.
- *Laboratory diagnosis*—the surface epithelium is destroyed and is replaced by a pseudomembranous meshwork of fibrin, necrotic epithelial cells, polymorphonuclear neutrophils and various types of microorganisms. The underlying connective tissue is markedly hyperemic with numerous engorged capillaries and a dense infiltration of PMN. Numerous plasma cells may appear in the periphery of infiltrate.

Differential Diagnosis

ANUG, presenting as an isolated phenomenon, must be differentiated from severe systemic diseases and in condition where acute inflammation of gingival and periodontal regions are the only associated symptoms.

- *Agranulocytosis*—blood picture shows evidence of a systemic disease and due to diminished immunity lesion is not marked by a severe inflammatory reaction.
- *Gingival changes due to cytostatic and immunosuppressive therapy*—history is relevant.
- *Benign mucus pemphigoid*—erosion, no necrosis and seen in elder adults.
- *Pemphigus*—it follows a chronic course with classical histological picture. It occurs in older patients and skin changes are seen.
- *Pemphigoid lichen planus*—no acute course, no bad breath, whitish changes and skin symptoms.
- *Gonococcal stomatitis*—the oral mucosa is covered with a grayish membrane that sloughs off in areas, to expose an underlying raw bleeding surface.
- *Syphilitic gingivitis*—primary lesion seldom on gingiva, does not spread to neighboring surfaces.
- *Gingivostomatitis due to Candida*—whitish deposits that can be wiped off, no bad breath, less acute picture and demonstration of fungi.
- *Tubercular ulcer*—less acute picture.
- *Streptococcal gingivostomatitis*—it is characterized by a diffuse erythema on the posterior areas of the oral mucosa. But in these cases, necrosis of gingiva is not a feature.

Management

- *Removal of pseudomembrane*—the involved areas are isolated with cotton rolls and dried. A topical anesthetic is applied and after 2 to 3 minutes, the areas are gently swabbed with a cotton pellet to remove the pseudomembrane and nonattached surface. After the area is cleansed with warm water, the superficial calculus is removed.

Table 24-3: Herpetic gingivostomatitis

ANUG	Acute herpetic gingivostomatitis
Etiology—interaction between host and bacteria, most probably fusospirochetal	Specific viral etiology
Necrotizing condition	Diffuse erythema
Punched out gingival margins, pseudomembrane that peels off leaving raw areas, marginal gingiva affected	Vesicle ruptures and leave a slight depression or an oval or a spherical ulcer
Rare in children	Occurs most frequently in children
No definite duration	Duration of 7 to 10 days

Table 24-4: Difference between ANUG, diphtheria and secondary stage of syphilis

ANUG	Diphtheria	Secondary stage of syphilis
Caused by fusospirochetes	Caused by corynebacterium diphtheriae	Caused by treponema pallidum
Affect marginal gingiva	Rarely affects the marginal gingiva. It affects the throat, fauces and tonsil	Rarely affects marginal gingiva. Any site of the oral cavity can be affected
Membrane removal easy	Membrane removal difficult	Membrane not detachable
Painful condition	Less painful	Minimal pain
Serological findings are normal	Serological findings are normal	Serological findings are abnormal
Doubtful contagiousity	Contagious	Only direct contact can cause this disease

- *Rinsing the mouth*—the patient is asked to rinse the mouth within every 2 hours, with a glassful of an equal mixture of warm water and 3% hydrogen peroxide. Twice daily rinse with 0.12% chlorhexidine are also effective.
- *Antibiotics*—patients with severe ANUG and lymphadenopathy are treated with antibiotics Penicillin V- 250 or 500 mg, 6 hourly or erythromycin—250 or 500 mg, 6 hourly with metronidazole 400 mg, 8 hourly, for 7 days are the drugs of choice.
- *Gingival curettage*—scaling is performed, if sensitivity permits. After the disease process is diminished, complete gingival curettage and root planning is done.
- *Supportive treatment*—supportive treatment consists of copious fluid consumption and administration of nutritional supplements.

Non-plaque-induced Gingival Disease

Bacterial Origin Gingival Disease

- **Mechanisms**—in rare occasions, when non-plaque related microorganisms override the host's innate immunity, infective gingivitis and stomatitis may occur in immunocompromised or non-immunocompromised individuals.
- **Organism responsible**—such infections may be caused by *Neisseria gonorrhoea*, *Treponema palladium*, *Streptococci*, *Mycobacterium chelonae* or other microorganisms.
- **Appearance**—the lesions manifest as fiery red, edematous, painful ulcerations, as asymptomatic chancres or mucus patches, or as atypical non-ulcerated, highly inflamed gingivitis. Biopsy supplemented by microbiologic examination reveals the underlying etiology.

Viral-induced Gingival Disease

Herpetic Gingivostomatitis

- **Mechanism**—herpes simplex virus has a low infection potential. Hence, after gaining entry into the oral mucosa it penetrates a neural ending and travels to trigeminal ganglion through smooth endoplasmic reticulum and remains latent for years. The virus has also been isolated from normal gingiva, gingivitis, acute necrotizing gingivitis and periodontitis cases.
- **Appearance**—the clinical features of primary herpetic gingivostomatitis include painful, severe gingivitis with redness, ulcerations with serofibrinous exudates and edema accompanied by stomatitis. In its initial stage, it is characterized by the presence of discrete spherical gray vesicles which may occur on the gingiva, labial and buccal mucosa; soft palate, pharynx, sublingual mucosa and tongue. After approximate 24 hours, the vesicles rupture and form painful small ulcers with a red elevated halo-like margin and a depressed yellowish or grayish-white central portion. This occurs in widely separated areas or in clusters where confluence occurs. Fever and lymphadenopathy are common. The incubation period is one week. Healing of ulcers occur spontaneously without scarring in 10-14 days. During the course of the disease, the generalized soreness of oral cavity interferes with eating and drinking.
- **Management**—the treatment includes careful plaque removal to limit bacterial superinfection, which delays the healing. In severe cases, including patients with immunodeficiency, systemic use of antiviral drugs such as acyclovir or valacyclovir is recommended.

Herpes Zoster

- **Organism**—varicella-zoster virus causes varicella (chickenpox) as the primary self-limiting infections which occur mainly in children and later activation of the virus in adults causes herpes zoster (shingles).
- **Appearance**—the intraoral ulcers usually involves gingiva, tongue and palate. The diagnosis is generally obvious due to unilateral occurrence of the lesions associated with severe pain. Healing of lesions take place in 1-2 weeks.
- **Management**—treatment consists of soft or liquid diet, rest, atraumatic removal of plaque and diluted chlorhexidine rinses. This may be supplemented by antiviral drug.

Gingival Disease of Fungal Origin

- **Candidiasis**—the most common intraoral fungal infection is candidiasis which is caused by *Candida albicans*, the proteinase-positive strain. It usually occurs as a consequence of reduced host defense, reduced salivary secretion, smoking and treatment with corticosteroids and broad-spectrum antibiotics. The most common clinical characteristics of gingival candidial infection is redness of the attached gingiva often associated with a granular surface. A diagnosis of the candidial infection can be done on the basis of culture, smear and biopsy. Topical treatment involves application of antifungal, nystatin, amphotericin B or miconazole.
- **Linear gingival erythema**—it is regarded as gingival manifestation of immunosuppression characterized by a distinct linear erythematous band limited to free gingiva. There is no attachment loss or pocket formation and the lesion does not respond to conventional plaque control methods and improved oral hygiene.
- **Histoplasmosis**—it is a granulomatous disease caused by *Histoplasma capsulatum*. The clinical manifestations include acute or chronic pulmonary histoplasmosis. The oral lesions are initiated as nodular or papillary and later may become ulcerative, painful with the loss of gingival tissue. They are sometimes granulomatous and the clinical appearance may resemble a malignant tumor. The diagnosis is based on clinical appearance, histopathology and/or culture.

Gingival Manifestations of Systemic Condition

- **Mucocutaneous disorders**—many mucocutaneous disorders may manifest intraorally in the form of desquamative lesions and ulcerations of gingiva (Fig. 24-31). The most important of these are lichen planus (Fig. 24-30), pemphigoid, pemphigus vulgaris, erythema multiforme and lupus erythematosus. Certain conditions



Fig. 24-30: Desquamative gingivitis present due to lichen planus.



Fig. 24-31: Ulceration of gingiva seen in desquamative type of gingivitis associated with dermatological disorders.

may also present themselves as desquamative gingivitis. These are factitious injuries, graft versus host disease, Wegener's granulomatosis, foreign body gingivitis, Kindler syndrome and squamous cell carcinoma.

- **Allergic reaction**—allergic reactions in the oral mucosa are uncommon. These reactions are type I (immediate type), which is mediated by IgE, or more often type IV (delayed type) mediated by T cells. The uncommon occurrence of these reactions intraorally may be due to the fact that much higher concentration of allergens is required for an allergic reaction to occur in the oral mucosa than in skin. The allergic reactions can be further classified on the basis of the type of allergen: Reactions due to dental restorative materials and reactions due to oral hygiene products, food and chewing gum.
- **Hematological disorders**—hematological disorders like leukemia may also manifest intraorally. Gingival manifestations in leukemia which includes extensive swelling, ulceration, petechiae and erythema are common in acute than in chronic forms. The pronounced gingival swelling is due to leukemic infiltration and

plaque-induced inflammation. Spontaneous gingival bleeding is also a common symptom of leukemic patients which develops secondary to thrombocytopenia. The primary concern is prevention of infection and bleeding. Careful and strict oral hygiene measures can reduce gingival inflammation. In some cases of iron deficiency anemia, gingival enlargement can occur.



Fig. 24-32: Enlargement of gingiva seen in iron deficiency anemia.

Gingival Recession

Types of Gingival Recession (By Miller)

- **Class I**—marginal tissue recession not extends to the mucogingival junction. There is no loss of interdental bone or soft tissue.
- **Class II**—marginal tissue recession that extends to or beyond the mucogingival junction, there is no loss of bone or soft tissue in the interdental area (Fig. 24-33).



Fig. 24-33: Class II gingival recession.

- **Class III**—marginal tissue recession that extend to or beyond the mucogingival junction in addition, there is loss bone and/or soft tissue in the interdental area or there is malpositioning of the teeth (Fig. 24-34).



Fig. 24-34: Class III gingival recession.

- *Class IV*—marginal tissue recession that extends to or beyond the mucogingival junction with severe loss of soft tissue or bone interdentally and/or severe malpositioning of the teeth (Fig. 24-35).



Fig. 24-35: Type IV gingival recession.

Traumatic Lesion of Gingiva

- *Chemical injury*—chemical injury due to various agents have been noticed. Chlorhexidine induced mucosal desquamation, acetylsalicylic acid burn (aspirin burn), cocaine burn, sloughing of mucosa due to dentifrice detergents, necrosis of gingiva due to use of para-formaldehyde are examples of it.
- *Physical injury*—physical injury to gingiva may occur due to heavy brushing forces; high abrasive of dentifrice and horizontal movement of brushing. Overzealous brushing can also produce damage to interdental papilla.
- *Factitious injury*—factitious injuries leading to gingival recession can produce because of picking at or

scratching of the gingiva with fingernail or pins, oral piercing or overzealous use of tooth pick.

- *Thermal injury*—extensive thermal burns may occur to oral mucosa especially palatal and labial mucosa due to hot beverages, pizza, melted cheese. The lesions may be painful, erythematous or with a slough coated surface. Vesicles may also form and sometimes may present as ulceration, petechiae or erosion.

Foreign Body Reaction

Another type of tissue reaction is observed when foreign materials are embedded in the gingival connective tissue due to epithelial ulceration. The common example is amalgam tattoo. Sometimes, chewing on sticks can introduce piece of stick in the gingiva producing an inflammatory reaction.

Carcinoma of the Gingiva

It is most commonly seen squamous cell carcinoma and occurs due to habits of tobacco.

Clinical Features

- *Age*—it is common in 5th and 6th decades of life.
- *Site*—the carcinoma of mandibular gingiva is more common than the involvement of the maxillary gingiva. The tumor arises most commonly in an edentulous area, although it may develop in a site where teeth are present. The fixed gingiva is involved more than the free gingiva. It usually occurs in premolar-molar area. In maxilla, gingival carcinoma often invades maxillary sinus or it may extend to palate or tonsillar pillars.
- *Appearance*—it is manifested as an area of ulceration, which may be a purely erosive lesion and may exhibit as an exophytic, granular or verrucous type of growth.
- *Symptoms*—in some patients, the first symptom may be loosening of the teeth.
- *Invasion*—quickly spreads from the gingiva to alveolar bone below. The proximity of the underlying periosteum and bone usually invites early invasion of these structures (Fig. 24-36).

Radiographic Features

- *Types*—there are, radiologically, three types of bone destruction, i.e. permeated type, moth eaten type and pressure type.
 - *Permeated type*—bone destruction with ill-defined margins. Permeated means permeation of water into sand, with ragged ill-defined borders. It also destroys the mandibular canal.
 - *Moth eaten type*—it resembles moth eaten image with remnants of bony fragments.



Fig. 24-36: Carcinoma of gingiva which is extending toward buccal mucosa (Courtesy Dr Suwas Darvekar).

- *Pressure type*—dish out concavity appearance with relatively smooth margins.
- *Rarefaction*—rarefaction with remnants of small bony fragment.
- *Cortical bone destruction*—cortical bone destruction with pathologic fractures also can occur.

Diagnosis

- *Clinical diagnosis*—ulcerative growth on the gingiva with history of tobacco habit may give clue to the diagnosis.
- *Radiological diagnosis*—comparison between periodontitis and carcinoma of gingiva is described below in **Table 24-5**.
- *Laboratory diagnosis*—biopsy show features suggestive of squamous cell carcinoma.

Table 24-5: Comparison between periodontitis and carcinoma of gingiva

Features	Periodontitis	Carcinoma of gingiva
Margin of bone resorption	Smooth and well defined	Infiltrative and ill-defined
Sclerotic bony findings	Exist, around the margin of the lesion	No sclerotic finding
Residual small bony fragments	None within the lesion	Exist in most of the cases
Size	The condition is usually generalized and if localized, there is a specific cause for it	Localized finding

Management

- *Surgery*—localized lesions, of less than 3 cm in diameter, were cured in 80% of the cases by surgical excision.

Periodontal Disease

Periodontitis is defined as an inflammatory disease of the supporting tissues of the teeth caused by specific microorganism resulting in progressive destruction of the periodontal ligament and alveolar bone with pocket formation, recession or both.

The clinical feature that differentiates periodontitis from gingivitis is presence of clinically detectable attachment loss. The clinical classification was simplified to describe the three general clinical manifestations of periodontitis: chronic periodontitis, aggressive periodontitis and periodontitis as a manifestation of systemic disease.

Etiology of Periodontal Diseases

- *Multifactorial*—multifactorial, resulting from various hosts and environmental factors.
- *Open contact*—when teeth are not in contact, the patient is said to have open contact. It is dangerous for the periodontium as food debris may trap in it and hence more bone loss occurs, than the areas of closed contact.
- *Microorganisms*—plaque forming bacteria play a role in initiation and progression of destruction of the periodontium, mainly the anaerobic bacteria. When bacteria colonize at the root surface, they spread in the region between the root and gingival margins and stimulate a chronic inflammatory reaction. It results in pocket formation and apical migration of the epithelial attachment and bone loss.
- *Immune system*—Neutrophils, granulocytes and monocytes are very important in causing destruction of the bacteria and thus, preventing periodontitis. It may, sometimes, release lymphocytes and cause microbial destruction.
- *Secondary local factors*—like calculus deposits (**Fig. 24-37**), carious cavities, overhanging margins, perforation by pins or posts, over erupted opposing teeth and gingivally impinging partial dentures.



Fig. 24-37: Heavy calculus seen in upper posterior region.

Pathogenesis of Periodontitis

It is an inflammation of the supporting structures of the teeth. It starts out as gingivitis that then spreads down to the root surface causing alveolar bone resorption and pocket formation. It can eventually lead to alveolar bone loss and damage to the periodontal ligament then can result in tooth loss. Other complications include periodontal abscess and osteomyelitis of the jaws.

Periodontal Pockets

Classification

- *Gingival pocket (relative or false)*—it is formed by gingival enlargement, without destruction of the underlying periodontal tissues. The sulcus is deepened because of the increased bulk of the gingiva (**Fig. 24-38**).
- *Periodontal pocket (absolute or true)*—there is destruction of the supporting periodontal tissue; progressive pocket deepening leads to destruction of the supporting periodontal tissues and loosening and exfoliation of the teeth.
- *Suprabony pocket (supracrestal or supra-alveolar)*—in it, bottom of the pocket is coronal to the underlying alveolar bone.
- *Infrabony (intrabony, subcrestal or intra-alveolar)*—in it, bottom of the pocket is apical to the level of the adjacent alveolar bone.



Fig. 24-38: Gingival pocket seen due to enlargement of gingiva.

Pathogenesis

- *Microorganism*—periodontal pockets are caused by microorganisms and their products, which produce pathologic tissue changes that lead to the deepening of the gingival sulcus.
- *Inflammatory changes*—pocket formation starts as an inflammatory changes in the connective tissue wall of the gingival sulcus.

- *Degeneration of connective tissue*—the cellular and fluid inflammatory exudates cause degeneration of the surrounding connective tissues, including the gingival fibers.
- *Destruction of collagen fibers*—just apical to the junctional epithelium, an area of destroyed collagen fibers develops and becomes occupied by inflammatory cells and edema. Collagen loss may occur due to enzymes, like collagenase and other lysosomal enzymes from polymorphonuclear leukocytes and macrophages, which become extracellular and destroy the collagen.
- *Proliferation of junctional epithelium*—as a consequence of loss of collagen, the apical portion of the junctional epithelium proliferates along the root, extending in finger-like projections.
- *Detachment of junctional epithelium*—as the apical portion migrates, the coronal portion of the junctional epithelium detaches from the root. As a result of inflammation, polymorphonuclear neutrophils invade the coronal end of junctional epithelium. An increase in number of these cells, results in loss of tissue cohesiveness and tissue detachment from the tooth surface. Thus, the bottom of the sulcus shifts apically, resulting in deepening of the periodontal pocket.

Clinical Features

- *Signs*—gingival bleeding or/and suppuration, tooth mobility and Diastema formation may be present. In some cases, pus may be expressed by applying digital pressure.
- *Symptoms*—patient may complain of localized pain or deep pain in the bone. When explored with a probe, the inner aspect of the periodontal pocket is generally painful.
- *Color*—there may be bluish red, thickened marginal gingiva and a bluish red vertical zone from the gingival margin to the alveolar mucosa.

Radiographic Findings

- *Use of gutta percha point*—pockets are not detected on radiograph as they are soft tissue changes. Gutta percha points or calibrated silver points can be used, with a radiograph, to assist in determining the level of attachment of the periodontal pocket.

Diagnosis

- *Clinical diagnosis*—this can be made with the help of periodontal probe (**Fig. 24-39**).
- *Radiological diagnosis*—it is made with the help of gutta percha point.
- *Laboratory diagnosis*—there may be circulatory stagnation, destruction of the gingival fibers and surrounding tissues, which results in discoloration.



Fig. 24-39: Periodontal pocket is examined with the help of periodontal probe.

Management

- *Pocket irrigation*—devices like squeeze bottles and blunt hypodermic needles can be used to irrigate the pocket with chemotherapeutic agents.
- *Flap surgery*—it is done to eliminate pockets.

Periodontal Abscess

It is usually culmination of a long period of chronic periodontitis.

Pathogenesis

- *Pre-existing periodontal pocket*—it usually occurs in pre-existing periodontal pocket. When such pocket reaches sufficient depth of about 5 to 8 mm, the soft tissues, around the neck of the tooth may approximate the tooth so tightly that orifice of the pocket is occluded.
- *Multiplication of bacteria*—bacteria multiply in the depth of pocket and cause sufficient irritation to form an acute abscess, with exudation of pus into this area. It results in sufficient swelling to destroy the cortical plates of bone.

Clinical Features

- *Location*—it starts at the gingival crevice and extends down on one or more surface of the root, frequently as far as apical region.
- *Symptoms*—acute episode usually has sudden onset with extreme pain. There is also distension and discomfort.
- *Signs*—they are associated with swelling of the soft tissues overlying the surface of the involved root (**Figs 24-40A and B**). Tooth is tender and mobile. Pus usually exudes from the gingival crevice.



Figs 24-40A and B: Periodontal abscess showing swelling at the periapical region.

Diagnosis

- *Clinical diagnosis*—severe pain with swelling in the apical region will give clue to the diagnosis.
- *Laboratory diagnosis*—there is central cavity filled with pus walled off on one side by the root and on the other side by connective tissue.

Management

- *Analgesic and antibiotics*—suitable analgesics and antibiotics should be given to patient.
- *Incision and drainage*—primary treatment for relief of acute symptoms is incision of the fluctuant abscess, from the depth of the abscess cavity to the gingiva. The incision should extend into the soft tissues of the root surface.
- *Debridement of root surface*—if the surrounding tissue are normal, the tooth may be retained and debridement of the root surface by removal of granulation tissue should be done. Treatment for new attachment and new tissue regeneration should be performed.

- *Extraction of tooth*—however, if the roots are denuded beyond the apical thirds of the root, the tooth should be extracted and curettage should be carried out to remove the granulation tissue from the socket.

Chronic Periodontitis

It is the most common form of periodontitis and is most prevalent in adults but can also be observed in children. It is generally a slowly progressive form of periodontitis which at any stage may undergo acute exacerbation with associated attachment loss. The clinical features are gingival inflammation, bleeding on probing, reduced resistance of the periodontal tissues to probing, loss of clinical attachment and loss of alveolar bone. Variable feature includes gingival enlargement, recession of gingiva, furcation involvement, increased tooth mobility, drifting of teeth and eventually exfoliation of teeth.

Chronic periodontitis is subject related with only a few individuals experiencing advanced destruction, affects specific teeth and the progression of inflammatory disease is continuous with brief episodes of localized exacerbation and occasional remission.

Classification

It is classified according to extent and severity of diseased sites

- *According to extent of disease:*
 - *Low*—1 to 10 sites.
 - *Medium*—11 to 20 sites
 - *High*—Greater than 20 sites.
- *According to severity*—this can be differentiated to the degree of clinical attachment loss.
 - *Mild*—1-2 mm clinical attachment loss.
 - *Moderate*—3-4 mm clinical attachment loss.
 - *Severe*—more than 5 mm clinical attachment loss (Fig. 24-41).



Fig. 24-41: Generalized severe chronic periodontitis.

Etiology

Many risk factors have been associated with initiation of chronic periodontitis or once contracted then the progression of the disease.

- *Microbial plaque*—it is a crucial factor in inflammation of the periodontitis is largely governed by host-based risk factors.
- *Age*—the prevalence of periodontitis increase with age as a result of cumulative effects of disease over a lifetime.
- *Smoking*—smoking has been positively associated with progression of periodontitis. The risk attributed to tobacco for chronic periodontitis is between 2.5 and 7.0. It is not only the risk for developing the disease, but also the response to periodontal therapy is impaired to smokers.
- *Host-related factors*—
 - *PMN*—reduction in number and/or function of PMNs.
 - *Drugs*—drugs like Phenytoin, nifedipine and cyclosporine predispose to gingival overgrowth and thus may modify preexisting chronic periodontitis.
 - *Hormones*—changes in circulating hormones during pregnancy, puberty, and menopause can modify gingival response to plaque.
 - *Immunosuppressant drug therapy*—immunosuppressant drug therapy and any disease resulting in suppression of inflammatory and immune processes (such as HIV infection) may predispose the individual to periodontal tissue destruction.
 - *Nutritional deficiency*—nutritional deficiencies in animals have shown effects on periodontal tissues but have been proven in human studies. Vitamin C deficiency has been consistently associated with increased gingival bleeding.
 - *Diabetes mellitus*—periodontitis severity goes hand in hand with poorly controlled diabetes mellitus. On the other hand, periodontitis may also exacerbate diabetes as it may decrease glycemic control.
 - *Psychogenic disorders*—stress and other psychosomatic disorders may have direct anti-inflammatory and/or immunosuppressive effects which are relevant to the etiology of chronic periodontitis and necrotizing ulcerative gingivitis.
 - *Genetic*—gingivitis is a general and ubiquitous response to plaque accumulation whereas risk of chronic periodontitis has a high inherited component. The focus of researcher is on the polymorphisms associated with genes involved in cytokine production. An increased risk for chronic periodontitis has been associated with polymorphisms of genes involved in cytokines production but these findings are yet to be confirmed.

Clinical Features

- *Age and sex*—it generally affects both the sexes equally and is seen in older age groups more frequently.
- *Site*—it is usually generalized, although some areas may be involved more deeply than others.
- *Symptoms*—it is usually painless, but sometimes exposed root may be sensitive to heat and cold, in absence of caries. Areas of localized deep pain, sometimes radiating deep into the jaws are established.
- *Signs*—the characteristic finding in it, is gingival inflammation, which results from accumulation of plaque and loss of periodontal attachment. The gingiva is slightly or moderately swollen and exhibits alteration in color from pale to magenta. There is also loss of stippling and gingival bleeding, which may be either spontaneous or easily provoked.
- *Pocket formation*—there is presence of pocket with variable pocket depth. Both, horizontal and angular bone loss can be found (Fig. 24-42).



Fig. 24-42: Chronic periodontitis showing pocket formation.

- *Tooth mobility*—tooth mobility is found in advanced cases, where bone loss has been considerable. Teeth give off a rather dull sound when tapped with a metal instrument. The embrasures may be opened because the interdental papillae are deficient.
- *Gingival recession*—gingival recession is a common phenomenon in later stages of disease, which may expose the cementum (Fig. 24-43).

Radiological Features

- *Bone resorption*—there is blunting of the alveolar crest due to beginning of bone resorption. Bone loss may occur horizontally, vertically (Fig. 24-44) or sometimes, in furcation area.



Fig. 24-43: Gingival recession seen in lower anterior region due to periodontal problems.



Fig. 24-44: Severe horizontal and vertical bone loss seen in chronic periodontitis.

- *Loss of interdental bone*—loss of corticated interdental crestal margin, the bone edges become irregular or blunted (Fig. 24-45).
- *PDL space widening*—widening of the periodontal ligament space and localized or generalized loss of alveolar supporting bone.
- *Floating teeth appearance*—in case of severe periodontitis, floating teeth appearance is seen (Fig. 24-46).

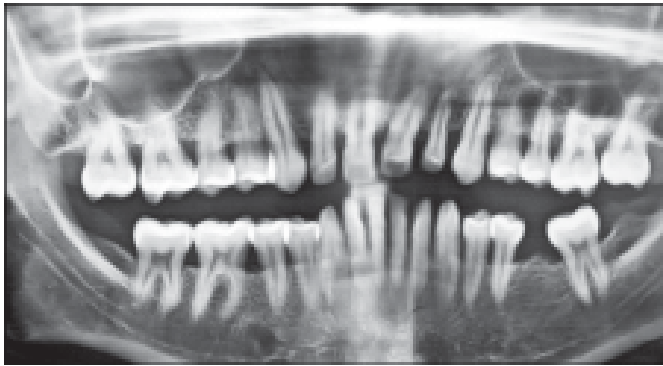


Fig. 24-45: OPG of chronic periodontitis showing bone loss.

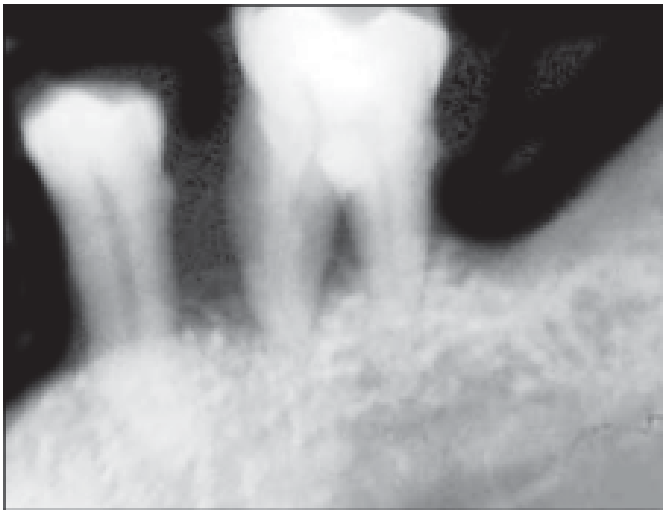


Fig. 24-46: Floating teeth appearance seen in periodontitis.

Diagnosis

- *Clinical diagnosis*—gingival recession, pocket formation with mobility of tooth will diagnose the condition.
- *Radiological diagnosis*—horizontal and vertical bone loss is present.
- *Laboratory diagnosis*—the enlarged free marginal gingiva is densely infiltrated with lymphocytes and plasma cells. The apical borders of the inflamed area approach the crest of the alveolar bone and the crestal fibers of the periodontal ligament.

Management

- *Oral prophylaxis*—scaling and curettage can be done.
- *Flap surgery*—this is the treatment of choice.

Aggressive Periodontitis

Aggressive Periodontitis is a group of diseases which was previously defined on the basis of age of onset and was hence named early onset periodontitis. But at the 1999 international classification workshop has clubbed a group

of rare, often severe, rapidly progressive forms of periodontitis, often characterized by early age of clinical manifestation and a distractive tendency for cases to aggregate in families, under a common heading of Aggressive Periodontitis.

Etiology and Pathogenesis

- *Virulent microbial infection*—the shorter duration of time over which a clinical manifestation of the disease is seen suggests a highly virulent microbial infection or a highly susceptible host, or a combination of both.
- *Microorganism*—many studies have demonstrated that the prevalence of *Actinobacillus actinomycetem comitans* predominantly in localized aggressive periodontitis (Previously called localized juvenile periodontitis) with other microorganisms such as *Capnocytophaga*, *Eikenella corrodens*, *Prevotella intermedia* and motile anaerobes such as *Campylobacter rectus*. Gram positive isolates of streptococci, peptostreptococci and Actinomycetes were also obtained. Generalized Aggressive periodontitis, formerly named as early-onset periodontitis and rapidly progressive periodontitis, have been frequently associated with the presence of *Porphyromonas gingivalis*, *Bacteroides forsythus* and *actinobacillus actinomycetem comitans*.
- *Host response*—an intense infiltration of polymorphonuclear neutrophils underlines the importance of these cells in the local defense against bacterial aggression and their potential role in host mediated tissue destruction. B-cell and plasma cells form the mononuclear defense cells. Plasma cell dominantly produce IgG and in less amount IgA. The IgG is actively present in GCF which forms a line of defense against initial bacterial invasion. Peripheral blood mononuclear cells of AgP patients may demonstrate reduced autologous mixed lymphocytes reactions, as well as a higher than normal response to B-cell mitogen. A high level of PGE2 and IL-1beta is observed in AgP patients. It has been demonstrated that T-helper cells are depressed in numbers compared to T-suppressor cell. Elevated level of antibodies specific to AgP-associated microorganisms have been found in crevicular fluid than in peripheral blood. An important findings in LAP and GAP patients have been decreased migration and function of PMNs. These abnormalities are usually minor in sense that they usually are not associated with infection other than periodontitis. These abnormalities in PMNs have been found to have familial aggregation. Other recent reports contradict suggest hyper-inflammatory state resulting in high levels of serum pro-inflammatory cytokines in AgP patients (Autosomal dominant).

Characteristic of Aggressive Periodontitis

- **Primary features**—aggressive Periodontitis was characterized by following major common features
 - *Medical history*—non-contributory medical history.
 - *Rapid loss*—rapid attachment loss and bone destruction.
 - *Familial aggregation*—familial aggregation of cases.
- **Secondary features**—these are considered to be general but not universally present are:
 - *Microbial deposit*—amount of microbial deposit inconsistent with the severity of periodontal tissue destruction.
 - *Levels of actinobacillus*—elevated levels of Actinobacillus Actinomycetem comitans and in some Far East populations, Porphyromonas gingivalis.
 - *Phagocytic abnormalities*—phagocytic abnormalities are common in this type of periodontitis.
 - *Hyper-responsive macrophage*—hyper-responsive macrophage phenotype, including elevated production of PGE2 and IL-1 beta in response to bacterial endotoxins.
 - *Self-arresting attachment loss*—progression of attachment loss and bone loss may be self-arresting.

Classification

Aggressive Periodontitis was sub-classified at the international workshop into localized and generalized forms. The following were the observed features:

- **Localized aggressive periodontitis (LAP):**
 - Circumpubertal onset.
 - Localized first molar/incisor presentation with interproximal attachment loss on at least two permanent teeth, one of which is first molar and involving no more than two teeth other than first molars and incisors.
 - Robust serum antibody response to infecting agents.
- **Generalized aggressive periodontitis (GAP) (Fig. 24-47):**
 - Usually affecting persons under 30 years of age, but patients may be older.
 - Generalized interproximal attachment loss affecting at least three permanent teeth other than first molar and incisors.
 - Pronounced episodic nature of the destruction of attachment and alveolar bone.
 - Poor serum antibody response to infecting agents.

It is important to underline that, in the present state of uncertainty regarding both the causative agents and the genetic and environmental susceptibility to Aggressive periodontitis; it is possible that LAP and GAP may simply represent phenotypic variations, of a single disease entity. Conversely, it is possible that different Aggressive periodontitis forms may manifest themselves with a



Fig. 24-47: Generalized type of aggressive periodontitis.

common clinical presentation. This aspect is of great diagnostic and therapeutic importance.

Clinical Features

- **Age**—the age of onset of the disease ranges from the middle to late teens and till 30 years of age
- **Site**—the lesions are more generalized and all or most of the teeth are affected, without any definite pattern of distribution.
- **Signs**—gingiva is acutely inflamed, often proliferated, ulcerated and fiery red (Figs 24-48A and B). Bleeding may occur spontaneously, or on slight provocation. In some cases, gingiva appears pink and free of inflammation; but in spite of this, deep pockets can be revealed on probing.
- **Systemic features**—some patients may have systemic manifestations like weight loss, mental depression and general malaise.

Diagnosis

- **Clinical diagnosis**—rapid loss of periodontal structure with systemic features may suspect this disease.

Management

- **Antibiotics**—if microflora contains gram-positive microorganisms, then it should be treated with 250 mg amoxicillin and 125 mg potassium clavulanate three times daily, for 14 days, along with scaling and root planning. If flora is gram-negative, then clindamycin should be given with dose of 150 mg, four times a day, for 7 days, along with scaling and root planning.

Prepubertal Periodontitis

It is a very rare condition which starts during, or immediately after the eruption of primary teeth. There are functional defects of neutrophils and monocytes. It is transmitted as an autosomal recessive trait. It is associated with Papillion-Lefevre syndrome.



Figs 24-48A and B: Aggressive periodontitis showing gingival recession and inflammation with loss of attachment.

- *Generalized form*—in the generalized form, inflammation is extremely acute and destruction of gingiva and alveolar bone is very rapid. Leukocytosis, otitis media, skin and upper respiratory infection may also be present. The disease involves all the primary teeth (**Fig. 24-49**) and may or may not involve the permanent dentition. It is refractory to antibiotic therapy.
- *Localized form*—in it, only few teeth are affected. Gingiva may appear normal. The progress of disease is slow. Otitis media is an infrequent finding. The disease responds to scaling and antibiotic therapy.

Necrotizing Ulcerative Periodontitis

It occurs after repeated, long-term episodes of necrotizing ulcerative gingivitis. It can be associated with AIDS. Inflammatory infiltrate in the lesions of ANUG can extend to the underlying bone, resulting in a deep crater like osseous lesion most often located in the interdental area



Fig. 24-49: Prepubertal periodontitis showing involvement of primary teeth.

(**Fig. 24-50**). There is a presence of deep interdental osseous crater but deep conventional pockets are not found, because the ulcerative and necrotizing character of the gingival lesion destroys the junctional epithelium the removing the mechanism of pocket deepening. Ulceration is frequent finding in this type of periodontitis.

As the patients have underlying predisposing factors, it should be treated in consultation with a physician. Medical evaluation and local, topical and systemic antimicrobial should be given, based on the result of laboratory tests.



Fig. 24-50: Ulcerative periodontitis present in lower anterior region.

Tooth Mobility

Tooth has a slight degree of physiologic mobility, which varies from different teeth and at different times of the day. Mobility beyond the physiologic range is termed as abnormal or pathologic mobility.

Causes

- *Trauma from occlusion*—it usually results in mobility of the teeth. It occurs initially as result of resorption of the cortical layer of bone leading to a reduced fiber support and later, to adaptation phenomenon, resulting in a widened periodontal space.
- *Loss of tooth support*—it can result in mobility. The amount of mobility depends upon the severity and distribution of bone loss.
- *Inflammation*—extension of inflammation from the gingiva or periapical region, into the periodontal ligament, may increase tooth mobility.
- *Periodontal surgery*—it temporarily increases tooth mobility for a short period.
- *Systemic disease*—some systemic diseases can cause increased tooth mobility, like in pregnancy due to physicochemical changes in the periodontal tissues.
- The destruction of surrounding alveolar bone, such as in osteomyelitis or jaw tumor, may also increase tooth mobility.

Radiographic Features

- *PDL space widening*—there is a widening of periodontal ligament space and a single root may develop an hourglass shape. In multi-rooted teeth widening of periodontal ligament space at apices and in region of furcation is seen.
- *Angular bone loss*—bone loss which surrounds the tooth is also present (**Fig. 24-51**).

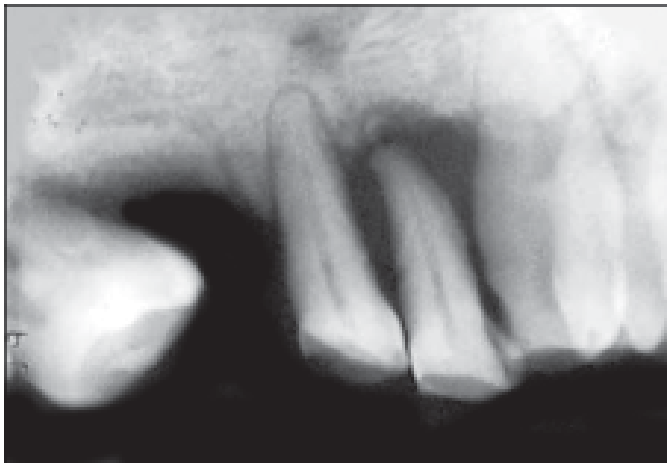


Fig. 24-51: Angular bone loss present in mobility.

Papillion-Lefevre Syndrome

It is an autosomal recessive and inherited disorder. It is a triad of:

- Hyperkeratosis of palms of the hand and soles of feet (**Fig. 24-52A**).

- Extensive prepubertal destruction of the periodontal bone supporting the dentition (**Fig. 24-52B**), usually extensive generalized horizontal bone loss.
- Calcification of dura.



Figs 24-52A and B: Papillion-Lefevre syndrome showing hyperkeratosis of palms with severe destruction of periodontium.

Clinical and Radiological Features

- *Hyperkeratosis*—there is reddened, scaly, rough palms and soles.
- *Periodontal finding*—It usually begins after eruption of the primary second molars. There is inflamed gingiva and horizontal bone destruction. Loss of entire primary dentition by the age of 5 years and loss of secondary dentition by the age of 20 years. Gingival swelling and severe halitosis.
- *Radiological features*—calcification of dura is seen in radiograph. Periodontal bone destruction is also present.

Diagnosis

- *Clinical diagnosis*—clinical diagnosis is suspected when hyperkeratosis of palm is seen in association with periodontal destruction.
- *Radiological diagnosis*—calcification in dura can be detected radiologically.
- *Laboratory diagnosis*—there is marked chronic inflammation of the lateral walls of the pocket, with predominantly plasma cell infiltrate, considerable osteoclastic activity and apparent lack of osteoblastic activity.

Management

- *Periodontal management*—flap surgery, oral prophylaxis should be done.

Radiological Assessment of Periodontal Diseases

Bone loss can be defined as a difference between the present septal bone height and the assumed normal bone height for any particular patient, bearing in mind that the normal bone height varies with age. The main radiographic projections used for periodontal diseases are bitewing, paralleling line angle periapical technique, digital radiography and dental panoramic tomography.

Advantages of Radiography in Periodontal Diseases

- *Identification of causative factors*—often valuable in identifying local initiating factors like calculus, poorly contoured or over extended restorations.
- *Status of periodontium*—to know the status of the periodontium such as root length and morphology, crown-root ratio, condition of the alveolar bone around the root and position of the maxillary sinus in relation to periodontal deformities.
- *Amount of bone present*—it offers the clinician to overview of the amount of bone present and serves as a permanent record of the condition of bone throughout the course of the disease.
- *Treatment outcome*—to evaluate treatment measures, particularly following guided tissue regeneration.

Limitations of Radiography

- *Overlapping*—inability of the viewer to perceive bony defects that are overlapped by existing bony walls.
- *Mild lesion*—earliest mild destructive lesions in the bone do not cause sufficient alteration in the density to be detectable.
- *Root superimposition*—density of the root superimposed with the image of the defect, tends to obscure bone height. Bone loss in furcation areas may be obscured by an overlying root or bony shadow.

- *Less accuracy of bone destruction*—radiographic image will tend to show less severe destruction than in the actual in the case.
- *Soft to hard tissue relationship*—soft to hard tissue relationship are not demonstrated on the radiographs.
- *Technique variation*—technique variation and X-ray beam position can considerably affect the appearance of the periodontal tissues.
- *Burn out*—overexposure can cause burn out.

Radiographic Features of Healthy Periodontium

- *Posterior region*—thin, smooth, evenly corticated margins of the interdental crestal bone in the posterior regions. It is just below the Cementoenamel junction. The interdental crestal bone is continuous with the lamina dura of the adjacent teeth. The junction of two forms a sharp angle (Fig. 24-53).
- *Anterior region*—thin, even, pointed margins to the interdental crestal bone in the anterior region. Cortication at the top of the crest is not always evident, mainly due to a small amount of bone between the anterior teeth. Thin and even width of the mesial and distal periodontal ligament space.

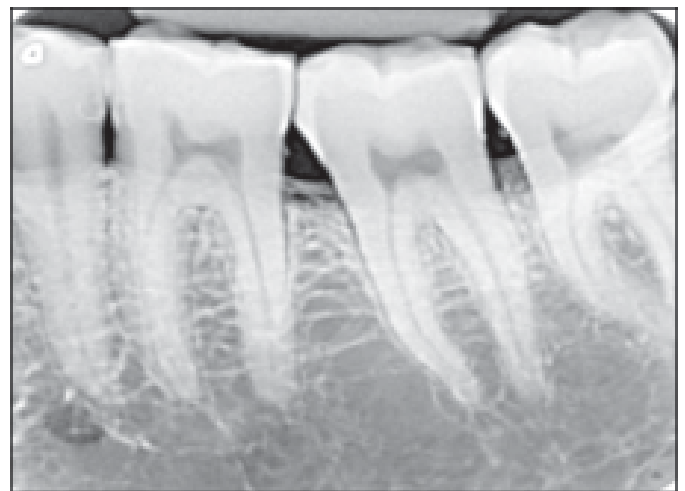


Fig. 24-53: Radiograph of normal periodontium.

Radiological Types of Periodontitis

Early Periodontitis

There are areas of localized erosion of the alveolar bone crest.

In the anterior region, there will be blunting of the alveolar bone. In some cases, it presents chisel edge, due to greater destruction of bone at the sides than at the summit.

In posterior areas there is loss of the sharp angle between the lamina dura and alveolar crest and it appears rounded off, with irregular and diffuse borders.

Moderate Periodontitis

There are two types:

Horizontal bone loss

- In horizontal bone loss, both, buccal and lingual bone plates and intervening inter-dental bone resorbed (**Fig. 24-54**). It does not indicate the current activity of disease.

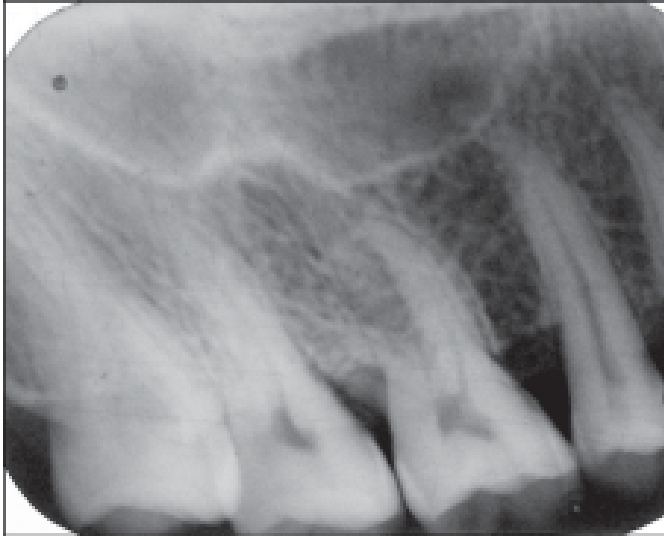


Fig. 24-54: Horizontal bone loss present between premolar and first molar.

- *Appearance*—radiographic appearance of loss in height of the interdental alveolar bone with the crest still horizontal and perpendicular to the long axis of the adjacent tooth.
- *Types*—it may be localized (**Fig. 24-55**) or generalized, depending on the areas involved. It may be mild, severe and moderate depending on the amount of bone loss.
- *Horizontal bone loss after surgery*—some patients, who have undergone periodontal surgery, may show

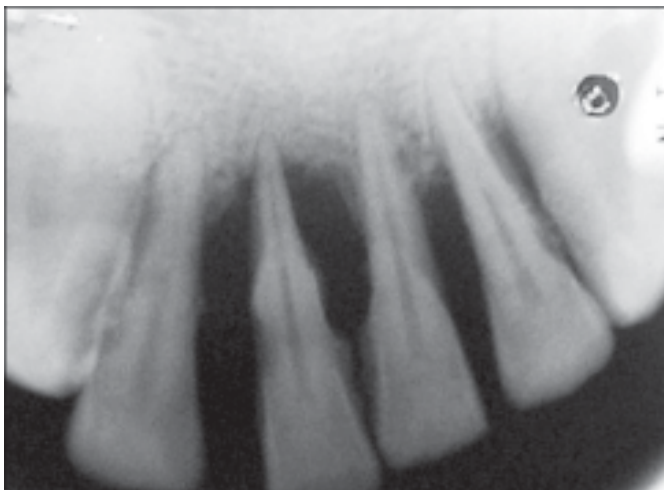


Fig. 24-55: Horizontal bone loss present in the anterior region.

moderate bone loss, but condition may be stable and without continuing bone loss and this may be indicated by a relatively dense apex.

Osseous defects

It is a type of bony lesion that may be seen in moderate periodontitis, other than horizontal bone loss. It is difficult to locate on radiographs. There are various types of osseous defects.

- *Inter-proximal crater*—it is the most common type of bony deformity. It is a trough like depression that occurs in the crest of inter-proximal septal bone between two adjacent teeth. It consists of two side walls vestibular and lingual cortical plates. The image of the side walls that is projected more coronally will be reduced in density, in relation to the image of apical side wall which is superimposed over the other side wall. If two walls are exactly superimposed, the inter-proximal crater may appear as an irregular linear area of reduced density between the adjacent teeth. Craters that are radiographically detected are about 1 mm or more in depth. The apical margin of the defect will not be sharply demarcated and will be such that relatively radiolucent image of the crater will gradually blend with the normal bone is apical to it.
- *Proximal intrabony defect*—it is a vertical defect within the bone. It extends apically, from the alveolar crest and is surrounded by three walls of bone, i.e. hemisepta and two marginal walls (lingual and vestibular) roots of the affected tooth is the fourth wall.
- *Hemisepta*—it is the bone of the interdental septum that remains on the roots of uninvolved adjacent tooth after destruction of either distal or mesial portion of the interproximal bone septum (**Fig. 24-56**). It occurs as a result of occlusal trauma which is complicated by inflammation. It is more common on distal surfaces of

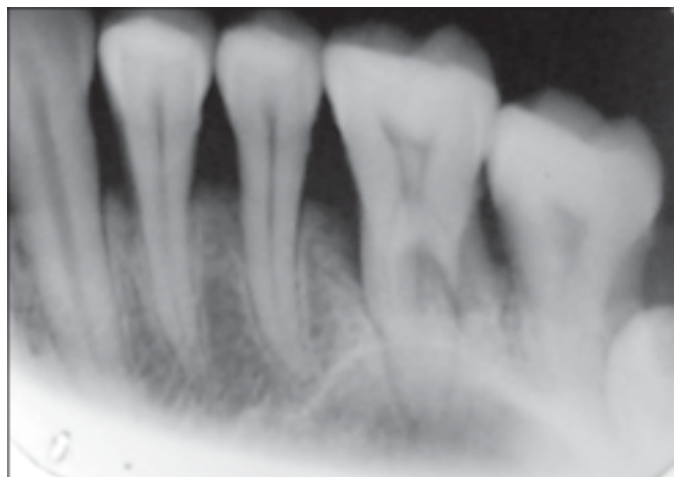


Fig. 24-56: Hemisepta present in premolar tooth.

teeth. Loss of attachment is increased in tooth mobility and open contact. It is V shaped and sharply outlined. Crestal lamina often appears at the coronal limit of the side wall. Gutta percha point is used to detect such cases.

- **Inconsistent bony margins**—it is the result of uneven resorption of alveolar cortical plates on the lingual or vestibular surface, with the result in that the crests of bony margins are irregular. It occurs when marginal bone is thin and removed by inflammatory process. It forms quite rapidly and remains constant for long periods. Difficult to identify on radiographs because it is obscured by the image of the root. Irregular margins apparent will relatively be opaque and more or less continuous with the lamina dura of interproximal crest, but apical to it. Radiographically it is not possible to determine the side of the tooth affected.
- **Bony pocket**—osseous defects, sometimes, form in the vestibular or lingual bone over the root and they are called as bony pockets (**Fig. 24-57**). They are surrounded by roots of involved teeth and cortical bone. They are extensions of proximal bony defect. Only detected when there is high degree of contrast between the thinned bone over the root and the surrounding bone.



Fig. 24-57: Bony pocket seen in relation with the mandibular first molar.

Advanced Periodontitis

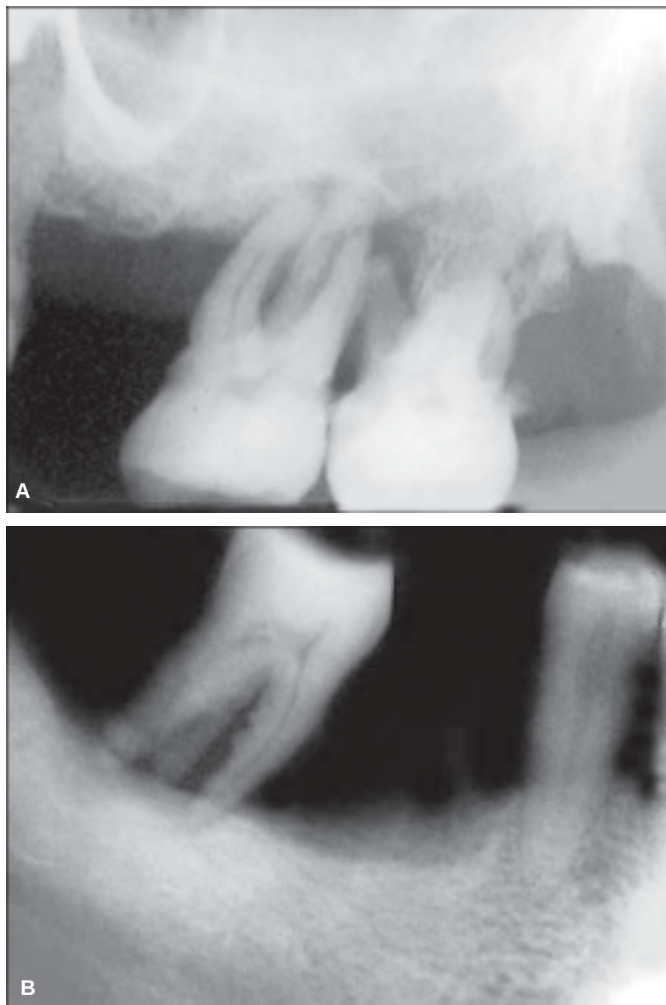
It is defined as the bone loss which is so extensive that the remaining teeth show excessive mobility, drifting and are in jeopardy of being lost due to inadequate support

Osseous Deformities in Furcation of Multi-Rooted Teeth

- **Furcation involvement**—progressive periodontal disease and its associated bone loss, may invade the bifurcation of multi-rooted teeth. It is found that the level of alveolar crest falls to occupy a position near the apices, so that

the major part of the root is often denuded. Bone resorption extends down the side of multi-rooted teeth, eliminating the marginal cortical bone over the root and it reaches the level of furcation and even beyond it.

- **Site**—the most common area for furcation involvement of maxillary 1st permanent molar is from its mesial side. Early maxillary molar furcation involvement between the mesiobuccal or distobuccal roots and the palatal root, produces a characteristic triangular shaped radiolucency at the edge of the tooth (**Figs 24-58A and B**). The pocket which is situated on the mesial or distal part of a tooth and occupies a small portion of the interdental septum on the lingual or buccal aspect only, may cross the adjacent root and involve the corresponding mesial or distal aspect of the bifurcation, producing only very slight bone changes.
- **Appearance**—mesial and distal furcation involvement is not apparent on radiographs because of presence of one or both cortical plates. If there is sufficient bone loss between lingual and buccal aspect of furcation in



Figs 24-58A and B: Severe bone loss seen in furcation area of molar teeth.

mandibular molar, radiolucent image will be sharply outlined. If it involves only one plate, then the image will be irregular in outline.

Alveolar Dehiscence

It results when the marginal bone chips apically and exposes the length of the root surface. The defect is wide and irregular and extends as far as the apex of affected teeth (**Fig. 24-59**). It occurs on either lingual or vestibular side of the tooth. On radiographs, it will appear as a faint radiopaque line representing its apical extension.

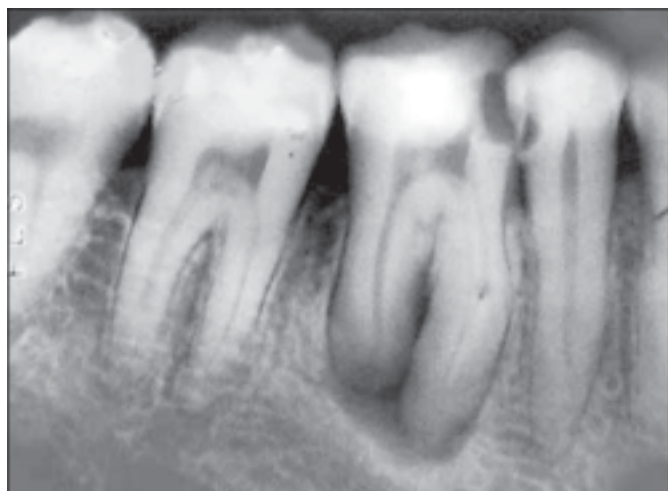


Fig. 24-59: Alveolar dehiscence showing faint line extending at the apex of tooth.

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Introduction

TMJ is a unique joint in which translatory as well as rotational movements are possible and where both the ends of bone articulate, in the same plane, with that of other bone. It is also called as ginglymodiarthrodial type of joint, meaning that it has a relatively sliding type of movement between bony surfaces, in addition to hinge movement, common to diarthrodial joint.

Anatomy of TMJ

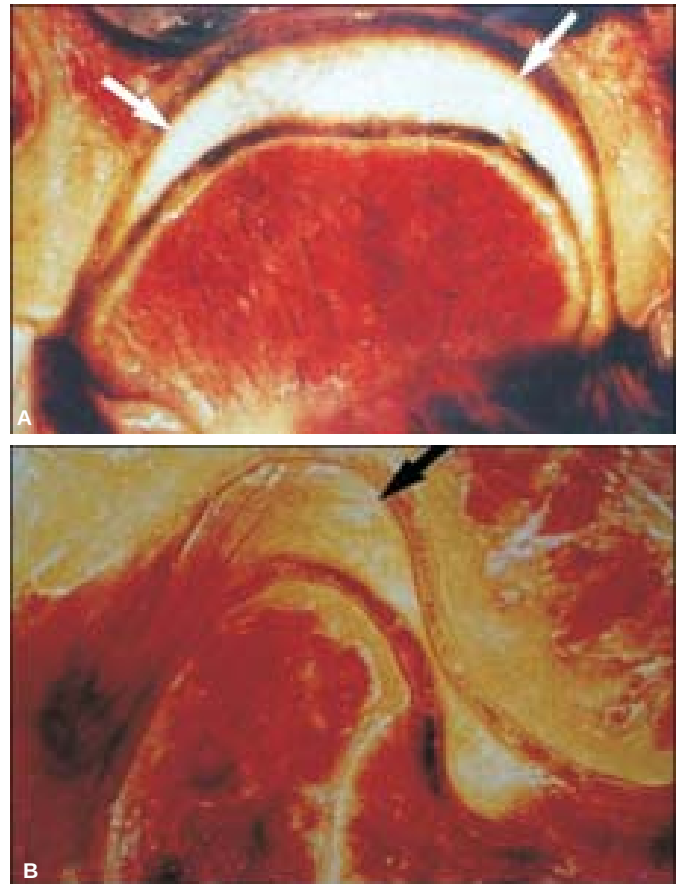
The TMJ is located between the mandibular fossa (glenoid fossa), the inferior surface of temporal bone and condylar process of the mandible. It is a synovial type of joint and it is distinguished from most of the joints by following points.

- **Fibrocartilage**—articulating surface of the bones is covered by avascular, fibrous connective tissue, which may contain variable number of cartilage cells. Thus called as fibrocartilage.
- **Point of closure**—the two articulating surface complex of bone carry teeth, whose shape and position influence the movement of joint. It is the only joint with rigid end point of closure.
- **Articulation**—it has bilateral articulation with cranium, so both the joints must function together. TMJ is a complex joint as it has an articular disc interposed between the condyle and the temporal bone (**Figs 25-1A and B**).

Joint Proper

Glenoid Fossa

The mandibular condyle articulates at the base of the cranium with the squamous portion of the temporal bone. This is called as the glenoid fossa.



Figs 25-1A and B: Normal positioning of disc in temporomandibular joint—front and lateral view.

- **Surface**—this is a continuous surface consisting of three regions:
 - The posterior slope to the height of convexity of the articular eminence just anterior to the glenoid fossa.
 - The flattened preglenoid plane continuing anteriorly from the height of eminence.
 - Entaglenoid process is continuous with the narrow glenoid plane.

- *Squamous tympanic fissure*—posterior to the glenoid fossa, there is a squamous tympanic fissure, which extends mediolaterally.
- *Post-glenoid process*—the posterior part of the fossa is elevated to form a ridge called the posterior articular ridge. This ridge increases in height laterally to form the thickened cone shaped prominence called the post-glenoid process, immediately anterior to the external acoustic meatus.
- *Entaglenoid process*—the lateral border of the fossa is usually raised to form a narrow crest joining the articular tubercle in front, with the post-glenoid process behind. Medially, the articular fossa narrows and is bounded by a bony wall, the entaglenoid process.
- *Roof*—the roof of the glenoid fossa is always thin and even translucent in many skulls. There is clear evidence that the articular fossa is not a stress bearing functional part of TMJ articulation.
- *Articular eminence*—the articular eminence is strongly convex in an anteroposterior direction and somewhat concave in a transverse direction. The degree of convexity varies, with the radius of the curvature varying from 5 to 15 mm.

Mandibular Condyle

- *Dimension and shape*—the condyle is about 15 to 20 mm long and 8-10 mm thick. Its long axis lies at right angle with the plane of the ramus. The condyle is usually quite convex anteroposteriorly and only slightly convex mediolaterally. The mediolateral convexity is often irregular, with medial and lateral slopes divided by a more or less prominent anteroposterior ridge.
- *Poles*—the articular surface of the mandible is seated on its ovoid condylar process. From the anterior view, it has medial and lateral projections called *poles*.
- *Lateral pole*—the lateral pole of condyle is roughened and often bluntly pointed.
- *Medial pole*—the medial pole is usually rounded and is more prominent than the lateral pole.
- *Lateral view*—in lateral view, the condyle appears tilted forward at the mandibular neck, with its articular surface on its anterosuperior aspect. The articular surface thus faces the posterior slope of the articular eminence, when the jaw is held with teeth in complete occlusion.
- *Articular surface*—the articular surface continues medially down and around the rounded medial pole of the condyle. Medial articular surface faces the entaglenoid process of the temporal bone, when the jaw is held in an occluded position.

Articular Disc

It is composed of dense fibrous connective tissue devoid of any blood vessels or nerve fibers.

- *Intermediate zone*—in the sagittal plane, it can be divided into three regions according to thickness. The central area is the thinnest and is called as *intermediate zone*. Both anterior and posterior to the intermediate zone, the disc becomes considerably thicker. The posterior border is generally slightly thicker than the anterior border.
- *Anterior view*—from the anterior view, the disc is generally thicker medially than laterally. The precise shape of the disc is determined by the morphology of the condyle and mandibular fossa. During movement, the disc is somewhat flexible and can adapt to the functional demands of the articular surface.
- *Retrodiscal tissue and lamina*—the articular disc is attached posteriorly to an area of loose connective tissue that is highly vascularized and innervated it is called as *retrodiscal tissue*. Superiorly, it is bordered by the lamina of connective tissue, which contains many elastic fibers, the superior *retrodiscal lamina*. This gives necessary freedom for anterior movement of the disc. Since this region consists of two areas, it is called as bilaminar zone.
- *Attachment*—the articular disc is attached to the capsular ligament anteriorly, posteriorly as well as medially and laterally. This divides the joint into two distinct cavities; the upper or superior cavity which is bordered by the mandibular fossa and superior surface of the disc and the lower or inferior cavity, which is bordered by the mandibular condyle and inferior surface of the disc.
- *Synovial fluid*—the internal surface of the cavity is surrounded by specialized endothelial cells that form the synovial lining. This lining along with a specialized synovial lining located at the anterior border of the retrodiscal tissue produce the *synovial fluid*, which fills both the joint cavities. Thus, TMJ is referred to as a synovial joint.

Ligamentous Structures

Functional Ligament

- *Collateral ligaments (Fig. 25-2)*—the collateral ligaments attach the medial and lateral borders of the articular disc to the poles of the condyle. It is commonly called as *discal ligament* and are two in number. The medial one attaches the medial edge of the disc to the medial pole of the condyle and the lateral one, attaches to the lateral edge of the disc to the lateral pole of the condyle. These ligaments are responsible for dividing the joint mediolaterally into the superior and inferior joint cavities. Their function is to restrict the movement of the

disc away from the condyle, as it glides anteriorly and posteriorly. The reason for it is that they contain collagenous connective tissue fibers which cannot be stretched. They have a vascular supply and are innervated. The innervations provide information regarding the joint position and movement.

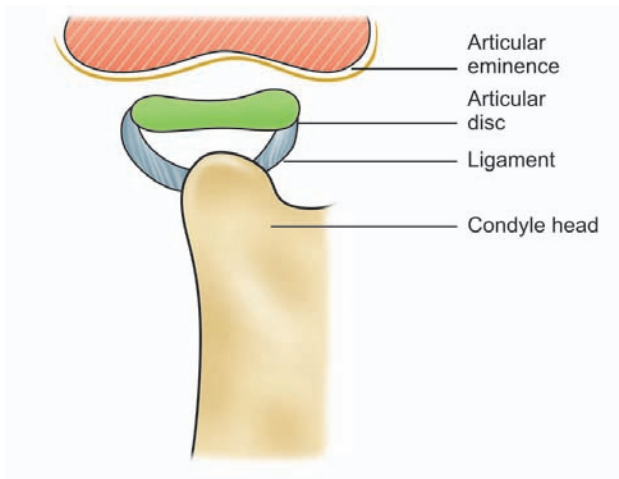


Fig. 25-2: Collateral ligament—A diagrammatic representation.

- **Capsular ligament (Fig. 25-3)**—the entire TMJ is surrounded and encompassed by the capsular ligament. The fibers of the capsular ligament are attached superiorly to the temporal bone, along the border of the articular surface of the mandibular fossa and articular eminence. Inferiorly, the fibers are attached to the neck of the condyle. It acts to resist any medial, lateral or inferior forces that tend to separate or dislocate the articular surface. Another function is to encompass the joint, thus retaining the synovial fluid. It is well innervated and provides proprioceptive feedback regarding the position and the movement of joint.

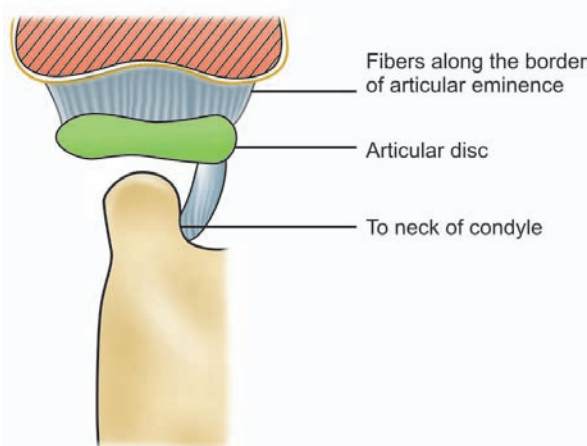


Fig. 25-3: Capsular ligament—A diagrammatic representation.

- **Temporomandibular ligament (Fig. 25-4)**—it is also called as *lateral ligament* as it is located laterally to the joint. It is composed of two parts, an outer oblique portion and an inner horizontal portion. The outer portion extends from the outer surface of the articular tubercle and zygomatic process, posteroinferiorly to the outer surface of the condylar neck. The inner horizontal portion extends from the outer surface of the articular tubercle and zygomatic process posteriorly and horizontally to the lateral pole of the condyle and posterior part of the articular disc. The oblique portion of the ligament resists excessive dropping of the condyle and therefore acts to limit the extent of mouth opening.

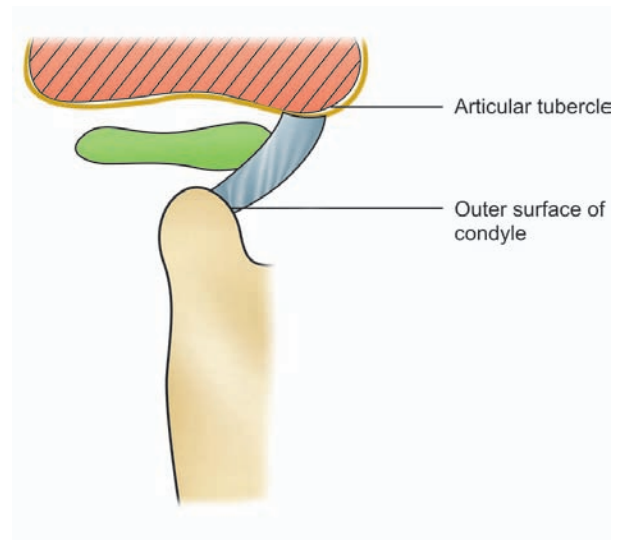


Fig. 25-4: Temporomandibular ligament—A diagrammatic representation.

Accessory Ligaments

- **Sphenomandibular ligament**—it is attached to the spine of the sphenoid bone and extends downwards and laterally to the small bony prominence on the medial surface of the ramus of the mandible, called the *lingula*. It does not have any significant effect on mandibular movement.
- **Stylomandibular ligament**—it arises from the styloid process and extends downward and forward to the angle and posterior border of the ramus of the mandible. It becomes taut when mandible is protruded but is most relaxed when the mandible is opened. Its function is to limit the excessive protrusive movements of mandible.
- **Mandibular malleolar ligament**—actually, the mandibular malleolar ligament consists of fibroelastic tissue with some ligamentous qualities. It originates from the neck and anterior process of malleus and is inserted on the medioposterior and superior part of the capsule, interarticular disc and sphenomandibular ligament.

Synovial Fluid

- **Volume**—the passive volume of upper and lower joint cavity is 1.2 and 0.8 ml respectively. A small amount of a clear, straw colored viscous fluid is found in the articular spaces, which is known as *synovial fluid*.
- **Source**—it is secreted by synovial membrane lining the articular disc, the capsule and also by retrodiscal tissue lining.
- **Properties**—synovial fluid is characterized by well defined physical properties of viscosity, elasticity and plasticity. It contains small population of varying cell type such as monocytes, lymphocytes, free synovial cells and occasionally polymorphonuclear leukocytes. The chemical composition of synovial fluid indicates that it is dialysate of plasma, with some added protein and mucin.
- **Gelation**—when the TMJ movements are reduced or restricted due to some reasons, the synovial fluid becomes viscid; its lubricating qualities are seriously impaired, a condition clinically referred to as *gelation*.
- **Functions of synovial fluid**
 - **Lubricant**—it is a lubricant and reduces the mechanical friction between the condyle and the articular disc and the mandibular fossa and articular disc.
 - **Nutritional fluid**—it is also a nutritional fluid for the vascular tissues covering the condyle and the articular tubercle and also for the disc. It is elaborated by diffusion from the rich capillary network of the synovial membrane, augmented by mucin secreted by the synovial cells.
 - **Liquid environment**—it provides liquid environment for the joint surface.
 - **Cleaning of joint**—synovial fluid is also considered to be responsible for the removal of entraneous material shed into the joint cavity. The intimate cells have been demonstrated to possess marked phagocytic properties.

Joint Innervations

- **Vascular supply**—it comes from the branches of the superficial temporal arteries, deep auricular arteries, anterior tympanic arteries and ascending pharyngeal arteries.
- **Nerve supply**—it is innervated by the branches of auriculotemporal nerve, masseteric nerve and the posterior deep temporal nerve, which are branches of the mandibular portion of the trigeminal nerve.

Functional Movement of TMJ

- **Elevation (jaw closing)**—the mandibular elevators include the coordinated functions of masseter, temporal and

medial pterygoid muscle of both the sides. Temporalis maintains the physiological rest position of the mandible. The posterior fibers of temporalis retract the head of mandible while closing the mouth.

- **Depression**—the depression of mandible includes the activity of the lateral pterygoid and the suprahyoid muscles. The inferior head of the lateral pterygoid is the main muscle used for depressing the mandible. The superior head of the lateral pterygoid pulls the articular disc forward, creating the glenoid joint activity. The suprahyoid group of muscles also acts in mandibular movement; by initiating and assisting opening of the jaw. The lateral pterygoid muscle has a major role, particularly when the mouth is opened wide or against resistance by the digastric, geniohyoid and mylohyoid muscle. The infrahyoid group of muscles participates in the activity by fixing the hyoid group to exert a downward pull on the mandible.
- **Protrusion**—it is performed by the medial and lateral pterygoid muscle of both the sides.
- **Retrusion**—it is performed by the posterior fibers of temporalis and digastric muscle.
- **Lateral excursive movements**—in this type of movement, the medial pterygoid and lateral pterygoid of each side, act alternately. If the mandible is moved to the right side the medial pterygoid of right side and the lateral pterygoid of left side act simultaneously.

Diagnostic Studies

Radiography

It is the most important diagnostic aid in distinguishing among the disorders that may affect the temporomandibular joint. Routine radiographic examination includes conventional radiography like Transcranial, Transorbital and Transpharyngeal view, etc. But main limitation is that it only shows osseous component and hence, the disease is diagnosed much later.

- **Computed tomography**—tomograms are superior to conventional radiography because of their ability to depict a greater portion of the joint. By providing the series of radiograph, tomography can reproduce small changes in the central portion of the TMJ and therefore decreases the false-negative interpretation (**Fig. 25-5**).
- **Arthrography**—with increasing interest in soft tissue derangement of the TMJ arthrography has assumed an important role in the diagnosis of joint disorders. Defect in the position or structure of the joint disc and its attachment can be determined using arthrography. Arthrography is performed by injecting the contrast media into the joint space and then after radiograph is taken.

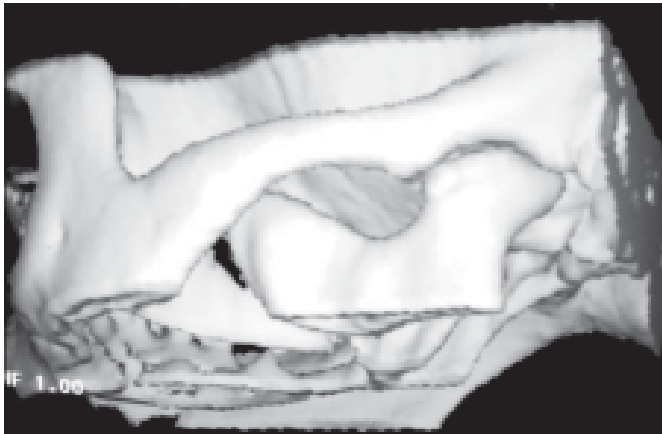


Fig. 25-5: 3D CT of temporomandibular joint showing condyle and glenoid fossa.

- *Electromyography*—it provides an objective means of providing monitoring the changes in the muscle activity and therefore can be a valuable aid in diagnosing MPDS and in evaluating the effectiveness of the treatment.
- *Arthroscopy*—steroid induced arthrography, chronic inflammatory changes in the synovium, acute inflammatory disease and gross damage to the condyle and disc could be visualized and diagnosed using arthroscopy in their study.

Table 25-1: Weldon Bell classification

Masticatory muscle disorders

- Protective muscle splitting
- Masticatory muscle spasm—MPD
- Masticatory muscle inflammation—myositis

Derangement of TMJ

- Incoordination
- Anterior disc displacement with reduction (clicking)
- Anterior disc displacement without reduction (mechanical restriction, closed lock)

Extrinsic trauma

- Traumatic arthritis
- Dislocation fracture
- Internal disc derangement
- Myositis
- Myospasm
- Tendonitis

Degenerative joint disease

- Non-inflammatory phase—arthrosis
- Inflammatory phase—osteoarthritis

Inflammatory joint disorders

- Rheumatoid arthritis
- Infective arthritis
- Metabolic arthritis

Chronic mandibular hypomobility

- Ankylosis—fibrous and osseous
- Fibrosis of articular capsule
- Contracture of elevator muscles
 - Myostatic contracture
 - Myofibrotic contracture
- Internal disc derangement—closed lock

Growth disorders of the joint

- Developmental disorders
- Acquired disorders
- Neoplastic disorders

Table 25-2: Second classification

Intracapsular

Degenerative joint diseases

- Osteoarthritis

Inflammatory

- Rheumatoid arthritis (and other collagen disorders)
- Psoriatic arthritis

Infection

- Gonorrhea
- Spread from contiguous sites
- Tuberculosis
- Syphilis

Developmental

- Condylar hyperplasia
- Condylar hypoplasia
- Agenesis

Traumatic

- Condylar fracture
- Ankylosis
- Dislocation
- Disc displacement

Metabolic

- Gout

Neoplasia

- Benign
- Malignant

Drug induced

- Steroid

Extracapsular

- Psycho-physiologic (MPD)
- Iatrogenic
- Traumatic
- Those referred from local dental origin
- Infection
- Otologic
- Neoplastic

Classification of TMJ Disorders

It is described in **Tables 25-1 and 25-2**.

Developmental Disorders of the TMJ

Hypoplasia of Condyle

It is the underdevelopment of mandibular condyle which can be congenital or acquired.

Causes

- *Prenatal growth disturbances*
 - *Hereditary*—hereditary anomalies like chromosomal anomalies, achondroplasia, mandibulofacial dysostosis, progeria, Larsen's syndrome and Goldenhar syndrome can lead to hypoplasia of condyle.
 - *Non-hereditary*—non-hereditary disease like Pierre Robin syndrome, radiation to fetus, Mobius syndrome are also causative factors.
- *Postnatal growth disturbances*
 - *Endocrine*—hypothyroidism and hypopituitarism causes decrease in secretion of growth hormone resulting in hypoplasia of condyle.

- **Trauma**—it is the most common cause of hypoplasia. Trauma may occur in infancy or childhood.
- **Nutritional factors**—deficiency of vitamin A and some other nutritional deficiency in the early infancy and childhood can lead to hypoplasia of condyle.
- **Infection**—rheumatoid arthritis can cause inhibition of growth of a component of the joint.
- **Irradiation**—radiation upon the face during the period of growth and extent of the change in the size of the condyle depends to some extent on the amount of radiation that is applied and the age at that time.

Clinical Features

Unilateral

- In this, only one side is involved. Following features can be present in unilateral hypoplasia.
 - **Appearance of face**—on the unaffected side, there is elongation of the body of mandible and flat appearance of the face. Body of mandible is short on affected side.
 - **Shifting on affected side**—mandible shift towards the affected side on opening.
 - **Malocclusion**—malocclusion is present. There may be cross bite on the affected side.
 - **Eruption of teeth**—eruption of teeth may be delayed in case of hypoplasia of the condyle. In some cases, it will cause impacted and unerupted teeth.
 - **Ear**—there may be deficiency of some parts of the adjacent auditory apparatus. The external ear may be small, deformed, partially or completely absent.

Bilateral

- In this, both the condyle are affected. Following features are present in this type.
 - **Mandible**—when there is bilateral arrest of condylar growth, there is usually symmetrical lack of growth of the mandible.
 - **Micrognathia**—due to lack of mandibular growth, micrognathia with the chin retracted to the level of hyoid bone occurs.
 - **Eruption**—there is delayed eruption of teeth on the both side.
 - **Class II malocclusion**—patient is having Class II malocclusion. It occurs as the mandibular ramus does not increase in height sufficiently to open the space between the upper and lower jaws into which the teeth erupt with concomitant growth of alveolar process. Another reason for this is that posterior growth of the ramus is affected, so the length of the body of the mandible is diminished and the last molars are left within the ramus.

Radiological Features

- **Condyle**—condylar process is short and it tends to assume a more posterior position in the glenoid fossa (**Fig. 25-6**). The neck of the condyle is slender.

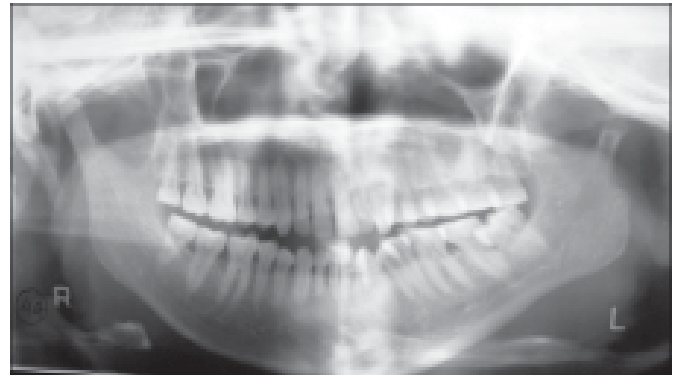


Fig. 25-6: Hypoplasia of condyle occurs on left side (Courtesy Dr Ashok L).

- **Ramus and body of mandible**—there may be proportionate shortening of the ramus and body on the affected side and the bone tends to be smaller than the opposite side. A shallow sigmoid and antegonial notch is also present.
- **Coronoid process**—coronoid process is relatively large, heavier and posteriorly directed.
- **Teeth**—teeth may be impacted.
- **Ear**—in some cases, there is congenital absence of the auditory canal and middle ear and the tympanic plate is poorly developed, so when the condyle is present, the articular fossa gives an appearance of an increased size.
- **Bilateral involvement**—in cases of bilateral underdevelopment, all the above features plus bilateral antegonial notching is seen.

Diagnosis

- **Clinical diagnosis**—malocclusion, elongation of body of mandible on unaffected side will give clue to the diagnosis.
- **Radiological diagnosis**—size of condyle is easily diagnosed on radiograph.

Management

- **Surgical**—surgical procedures commonly used in unilateral deformities are directed towards contributing bulk and increasing the length by means of bone, cartilage and soft tissue grafts. Most commonly graft is costochondral rib graft.
- **Orthodontic treatment**—it is done to correct malocclusion.

Agenesis of the Condyle

It is rare disorder. It is seen in hemifacial macrostomia, Goldenhar syndrome and Hallermann-Streiff syndrome.

Clinical Features

- *Site*—it can occur unilaterally or bilaterally and is a very rare condition.
- *Symptoms*—there are free movement (eccentric movement), anterior open bite, asymmetry of face, altered occlusion. Mastication may be difficult.
- *Signs*—shift of mandible towards the affected side occurs during opening in unilateral type, but it is absent in bilateral type.
- *Associated anomalies*—it is frequently associated with other anomalies like defective and absent external ear, an underdeveloped mandibular ramus or macrostomia.

Radiological Features

- *Absence of condyle*—absence of condyle is easily detected on radiograph.

Diagnosis

- *Clinical diagnosis*—eccentric movement with asymmetry of face.
- *Radiological features*—absence of condyle can be seen radiologically.

Management

- *Maintenance of dental health*—dental intervention can aid by establishment of an acceptable plane of occlusion, maintaining the oral health and offering palliative treatment for discomfort.
- *Osteoplasty*—if the derangements are severe, osteoplasty is advocated.

Hyperplasia of the Condyle

It is rare disorder which is characterized by excessive growth of condyles.

Causes

- *Developmental*—it may occur due to trauma during the birth.
- *Neoplastic*—some tumors like chondroma, osteochondroma or osteoma of condyle can lead to hyperplasia of condyle.
- *Bone disease*—fibrous dysplasia, Paget's disease which affects the bones, may affect the condyle leading to its overgrowth.
- *Hereditary*—this is also suggested the cause for condylar hyperplasia. Condylar hyperplasia can be seen in hereditary syndrome like Klinefelter syndrome.
- *Endocrine*—endocrine disorders like gigantisms as it affects the whole body, it can also affect the condyle causing its overgrowth.

- *Hypertrophic arthritis*—this is rare condition and it may lead to enlargement of the mandibular condyle.
- *Local circulatory disturbance*—local circulatory disturbance around the condyle may activate bone formation in that area causing its overgrowth.

Clinical Features

- *Age and sex distribution*—it occurs in either sex and there is no predilection for any side. Most common age of occurrence is 15 to 19 years.
- *Unilateral type*—it can affect only one condyle. Following features are present in unilateral type:
 - *Mandibular enlargement*—there is progressive enlargement of the mandible on the affected side.
 - *Facial asymmetry*—enlargement on the affected side, give rise to facial asymmetry on that side.
 - *Shift in midline*—due to facial asymmetry, shifting of the midline of chin to the unaffected side.
 - *Malocclusion*—unilateral type will result in cross bite, open bite, and asymmetric protrusion on the affected side.
- *Bilateral*—in this both the condyle are affected. Following features are present in bilateral type:
 - *Anterior cross bite*—mandible is larger on both side and is placed more forward than the maxilla. This will result in anterior cross bite (mandibular teeth are anterior to the maxillary teeth).
 - *Signs*—obtuse mandibular angle and the sigmoid notch form an arc of a larger circle of mandible.
 - *Relative microdontia*—as there is definite disproportion between normal size of crown of teeth and larger size of jaw bones, teeth appear to be smaller as compared to large size of jaw.

Radiological Features

- *Ramus*—the vertical ramus is increased in vertical depth as well as in its anteroposterior diameter. It will result in prevention of occlusion of the posterior teeth.
- *Body of mandible*—body of the affected side of mandible are larger as compared on unaffected side.
- *Condyle*—the condylar enlargement is sometimes symmetrically distributed throughout the whole process. It may retain its normal shape or it may assume a conical, spherical, pear shaped or an uneven and lobulated shape (**Figs 25-7 and 25-8**). The neck of the condyle may retain its integrity, be enlarged or absorbed into the enlarged head of the condyle.
- *Articular eminence*—the articular eminence is shallower than the opposite normal side, with the distal surface slightly evacuated.
- *Displacement of condyle*—hyperplasia of condyle may result in displacement of condyle from the mandibular fossa.



Fig. 25-7: Large size of condyle seen on the computed tomogram (Courtesy Dr Avinash Kshar and Dr Umarji).

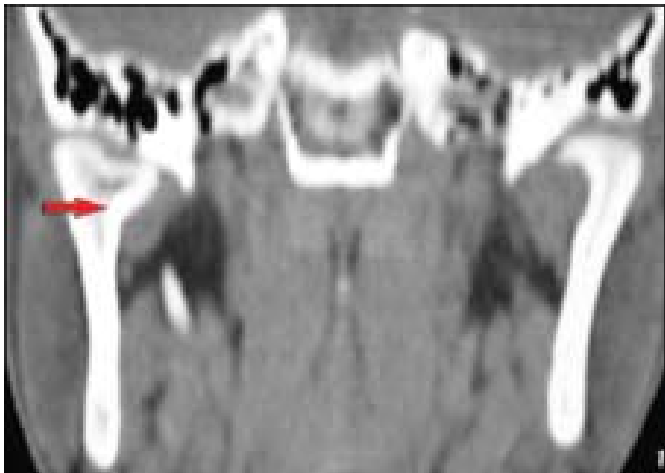


Fig. 25-8: Note large size of condyle on right side (arrow) as compared to on left side (Courtesy Dr Avinash Kshar and Dr Umarji).

- **Mandibular angle**—the angle of the jaw is right angle in some cases and it may be more obtuse than normal in some cases.
- **To assess the degree of bone activity**—it is done with the help of Scintigraphy using ^{99m}Tc .

Diagnosis

- **Clinical diagnosis**—enlargement of mandible with shift in midline will give clue to diagnosis.
- **Radiological diagnosis**—enlargement of condyle can be seen radiologically.

Differential Diagnosis

- **Hemifacial hyperplasia**—it is associated with soft tissue and teeth enlargement.

Management

- **Surgery**—unilateral condylectomy should be performed to improve function and esthetics of the patient.
- **Maxillary osteotomy**—some time, there may be occurrence of compensatory maxillary growth. In these cases, maxillary osteotomy should be performed.
- **Orthodontic therapy**—this is done to treat cross bite of the patient.

Double Condyle or Bifid Condyle

In this, double headed mandibular condyle is seen. Condyle is divided by an anteroposterior groove.

Cause

- **Trauma**—childhood fracture may lead to bifid condyle.
- **Developmental**—persistence of the well vascularized fibrous tissue septa, which is normally present in the condylar cartilage during embryonal and early postnatal life. Possible rupture of some of the blood vessels contained within the septa might impair the ossification of the condyle so as to cause a bifid development of the condylar head.
- **Abnormal muscle attachment**—this will also cause bifid condyle.

Clinical Features

- **Age and sex distribution**—it is noticed at any age and is more common in females than male with a ratio of 3:2.
- **Location**—it is usually unilateral. But bilateral occurrence can also be seen.
- **Symptoms**—limitation of opening of mouth, a small lateral deviation. In some cases, click of pop is heard while patient open the mouth.
- **Signs**—lateral movement of mandible is limited.

Radiological Features

- **Bilobed appearance**—radiographs will show bilobed appearance of the condyle (**Fig. 25-9**).
- **Separate glenoid fossa**—there may be two separate glenoid fossa.

Diagnosis

- **Clinical diagnosis**—not so specific.
- **Radiological diagnosis**—bilobed appearance can be seen.



Fig. 25-9: Bilobed appearance seen on radiograph of bifid condyle.

Management

- **Surgical**—as such no treatment is necessary, but if it is causing some problems surgical approach is carried out.

Coronoid Hyperplasia

It is rare developmental disorder affecting the coronoid process of mandible.

Causes

- **Endocrine**—as most of the cases are seen in puberty in the males, some sort of endocrine influence is suggested as the cause of coronoid hyperplasia.
- **Hereditary**—some cases are reported in sibling as hereditary factors may also play some role in coronoid hyperplasia.
- **Tumors**—tumors like osteoma and osteochondroma can also cause unilateral enlargement of the coronoid.

Clinical Features

- **Age and sex distribution**—it is seen in 2nd decade of life and most commonly encounter in males in the ratio of 5:1.
- **Unilateral coronoid hyperplasia**—in unilateral type, there is restriction of mandibular movement. It occurs due to enlarged coronoid may impinge on the zygomatic surface causing reduction in the mandibular movement. There is also deviation of mandible on the affected side.
- **Bilateral coronoid hyperplasia**—in this, limitation of movements occur which become more severe as the age progress.

Radiological Features

- **Unilateral**—In this case irregular nodular growth of the tip of the coronoid process is seen.
- **Bilateral**—there is symmetrical enlargement of coronoid on both sides.

Diagnosis

- **Clinical diagnosis**—not so specific.
- **Radiological diagnosis**—coronoid growth can be seen radiologically.

Management

- **Coronoidectomy**—it is carried out to remove the elongated process of coronoid.

Degenerative Joint Diseases

Osteoarthritis

It is also called as 'osteoarthrosis' or 'degenerative arthritis'. It is primarily a disorder of movable joints characterized by deterioration and abrasion of the articular cartilage with formation of new bone at the joint surface. There is destruction of the soft tissue component of the joint and subsequent erosion with hypertrophic changes in bone. There is breakdown of the connective tissue covering the condyle, articular eminence and the disc. Recently, there are some evidences suggesting that there are some inflammatory components present in osteoarthritis.

Etiopathogenesis (Fig. 25-10)

- **Overload to joint**—the lesion is brought by an increase in the functional demands of the healthy tissue due to repetitive overload on joint. This will result in breakdown of the joint.

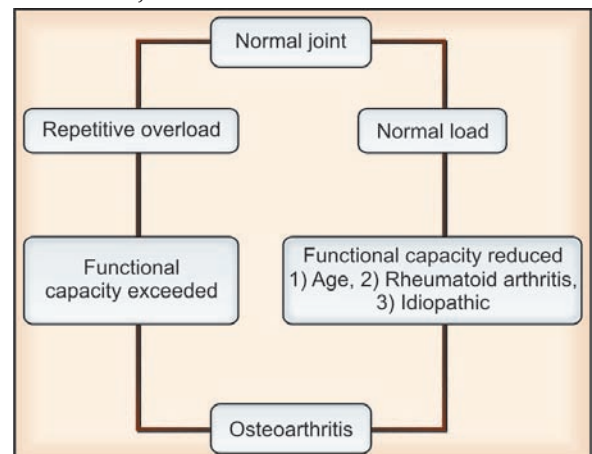


Fig. 25-10: Diagrammatic representation of etiopathogenesis of osteoarthritis of TMJ.

- *Deterioration of functional capacity of joint*—there may be normal load to the joint but functional capacity is reduced as a part of aging. This occurs due to:
 - *Slower replacement of chondroblasts*—as age advances there is slower replacement of chondroblasts and chondrocytes in the joint.
 - *Susceptible fibers*—the cartilage matrix turns over less rapidly resulting in available fibers to work for longer period of time. This will make them susceptible to fatigue.
 - *Poor nutrition to joint*—as matrix contain less water, the marrow blood flow diminished which results in poor nutrition to the joint. This will make joint desiccated and brittle.
- *Remodelling theory*—by another theory, bone growth does not cease completely after puberty and remodelling of the joint progresses under functional demands. Degenerative joint disease may develop when the remodelling rate of bone exceeds that of the cartilaginous repair. The gross evidence of these changes is the formation of marginal osteophytes with development of new bone in the area adjacent to the cartilage.

Causes of Secondary Osteoarthritis

- *Developmental*—Perthes disease, epiphysiolysis.
- *Metabolic*—alkaptonuria, hemochromatosis. Wilson's disease and chondrocalcinosis.
- *Traumatic*—intra-articular fracture, menisectomy, hypermobility, long leg arthropathy.
- *Endocrine*—acromegaly, gout.
- *Others*—hemophilia, aseptic necrosis, sickle cell disease, decompression sickness, tabes dorsalis etc.

Types

- *Primary*—it is generally described as a condition due to wear and tear and is more common with increasing age. It is commonly seen in patients more than 50 years of age.
- *Secondary*—the joint changes occur in response to recognizable local or systemic factors. It is more commonly seen in young persons.

Clinical Features

- *Age and sex*—it occurs in patients older than 40 years of age and 85% of them are older than 70, with a mean age of 53 years. Females are affected 6 times as frequently as males.
- *Joints involved*—it is common in many joints, but it is not frequently found in TMJ.
- *Symptoms*—there is unilateral pain over the joint, which may be sensitive to palpation. Patient also experience

pain on movements or biting, which may limit mandibular function. Pain usually worse in the evening.

- *Signs*—there is deviation of the jaw towards the affected side. Affected joint is swollen and warm to touch. Stiffness of the joint.
- *Crepitations*—there is presence of crepitation of the joint, the sound indicates degeneration within the articulating surfaces of the joint or disc.
- *Jaw movement*—there is limitation of jaw movements, which becomes increasingly apparent with function. Pain is usually located to the immediate preauricular region.
- *Spasm of muscle*—early signs may progress to spasm of the masticatory muscles resulting in stiffness and locking of the jaw. If not treated at this stage, it may lead to irreversible changes in the TMJ.
- *Course*—normal course is between 1 to 3 years. Severe symptoms last for about nine months, but gradually burn out leaving little or no disability.

Radiographic Features

- *Location*—degenerative changes located on the lateral and anterolateral wall of the fossa.
- *Erosion of condyle*—first evidence of erosion of condyle on a radiograph occurs on an average, 6 months after the onset of TMJ pain. This will result in enlargement and shallowing of mandibular fossa (**Fig. 25-11**).

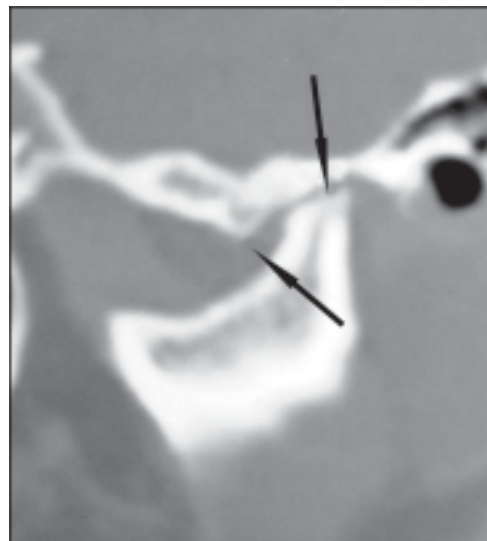


Fig. 25-11: Erosion of condyle seen in osteoarthritis of TMJ (Courtesy Dr Avinash Kshar).

- *Sclerosis*—density is increased as a result of sclerosis. Small crescent-like excavation appears at the superior aspect of the condyle just behind the point of articular contact. This is followed by wooly appearance, with spreading of rarefaction in the bone beneath the articular surface.

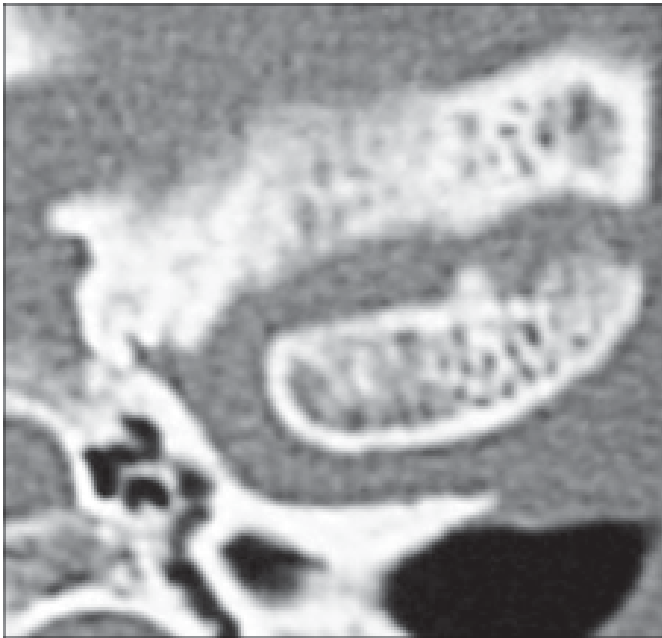


Fig. 25-12: Osteophyte formation seen as radiopaque projection on CT scan (Courtesy Dr Avinash Kshar).

- *Saucer shaped lesion*—fully developed lesions are saucer shaped on PA view. This is also called as the destructive phase.
- *Flattened articular eminence*—eminence is flattened or almost removed and anterior half of the superior convex surface of the condyle is converted into a flat plane.
- *Eburnation*—subchondral sclerosis of the condylar head becomes more dense and more radiopaque, is sometimes referred as eburnation.
- *Lipping*—development of lipping (shell like extension) on the anterior borders.
- *Osteophyte formation*—little shreds of the tissue at the margins of the articular cartilage surface may undergo ossification, so that small bony outgrowths or spurs develop which are called as osteophyte (**Fig. 25-12**).
- *Beaking*—extensive osteophytic formation is referred as *beaking*. These usually appear on the anterosuperior aspect of the condyle and lateral aspect of temporal component. In some cases, there is formation of sharp angle, either at the margins or actually on the surface of the articular process.
- *Joint mice*—osteophytes may break off and lie within the joint space, these fragments are called as '*joint mice*'.
- *Loose body*—osteophytes may be separated from its attachment and lie loose in the joint as a type of '*loose body*'.
- *Ely's cyst (subchondral cyst)* (**Figs 25-13 and 25-14**)—minute areas of degeneration filled with fibrous tissue are seen just below the bony surface of the condyle. The small radiolucent areas are usually less sharply defined and may have slightly irregular borders. Some are surrounded by an area of increased density, which may



Fig. 25-13: Ely's cyst presented as radiolucent area in the condyle.

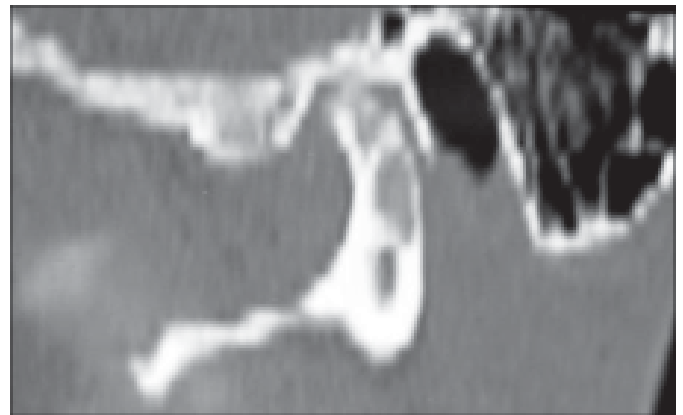


Fig. 25-14: Ely's cyst seen on CT scan in condylar region (Courtesy Dr Avinash Kshar).

be thin and well defined or relatively wide and not so sharply defined. These areas are regarded as cystic and are given the name '*Ely's cyst*'.

- *Joint space*—narrowing of the joint space and bony ankylosis may be seen.
- *Severe cases*—in severe cases, glenoid fossa may appear grossly enlarged because of the erosion of the posterior slope of articular eminence. The condyle may be markedly diminished in size and altered in shape due to erosion and destruction of the condylar head.

Diagnosis

- *Clinical diagnosis*—swollen joint in elderly patient with limitation of jaw movement may give clue to the diagnosis.
- *Radiological diagnosis*—Ely's cyst, osteophytes formation, joint mice, sclerosis, condylar erosion are typical of osteoarthritis
- *Laboratory diagnosis*—biopsy shows cells clumping together and become unevenly distributed throughout the fibrous matrix. It will also show loss of articular surface.

Management

- *Elimination of the cause*—it includes occlusal adjustment or replacement of the missing teeth and ill fitting prosthesis, grinding, treatment of caries and periodontal disease.
- *Relieving the pressure on joint*—occlusal adjustment and occlusal splints may reduce pressure on joint and relieve the symptoms.
- *Analgesic and anti-inflammatory drugs*—for the relief of pain, nonsteroidal anti-inflammatory drugs and analgesics should be given.
- *Physiotherapy*—heat therapy, diathermy and ultrasonic.
- *Myotherapy*—muscle exercises, injection of local anesthetic in TMJ.
- *Arthroscopic lavage*—arthroscopic lavage may give relief in some patients.
- *Doxycycline*—nowadays, low dose doxycycline (collagenase inhibitor, anti matrix metalloproteinase) is giving relief in many patients.
- *Other therapy*—glucosamine, chondroitin sulfate have also shown some success in osteoarthritis of TMJ.

Inflammatory Disorders of the Joint

Rheumatoid Arthritis

It is a debilitating systemic disease of unknown origin, characterized by progressive involvement of the joint, particularly bilateral involvement of large joints. Bony components of the TMJ are affected secondary to the granulomatous involvement of the synovial membrane that subsequently spreads to the articular surface of the condyle. It is non-suppurative inflammatory destruction of joint.

Etiopathogenesis

- *Phase one*—it result from some systemic infection, which evokes an inflammatory response within the joint.
- *Phase two*—as an autoimmune reaction to the antigen generated by the initial inflammation itself or it may be associated with derangement of the immune response to the exogenous antigen.
- *Active phase*—in the *active phase*, TMJ may get involved bilaterally. Bony components of the TMJ are affected secondary to the granulomatous involvement of its synovial membrane that subsequently spreads to the articular surface of the condyle. The joint space enlarges with synovial effusion which attacks the fibrocartilage and ultimately produces erosion of the underlying bone. This causes pain, stiffness and limitation of movement.
- *Chronic phase*—*chronic phase* may follow after active phase. Here, there is proliferation of the synovial membrane due to inflammation this is called as *pannus formation*. This pannus then encroaches the joint space and causes destruction of the articular cartilage. In this

lipping of the condyle and marginal proliferation is seen, this result in narrowing of joint space. Here, predominant clinical findings are crepitus, pain on biting and tenderness. The granulomatous tissue replaces the articular surface and small adhesions develop between the articular surface and disc.

- *Healing phase*—the process then enters the *healing* phase, where the symptoms subside and remodelling of the articular surface occurs.
- *Psychosomatic*—emotional trauma, anxiety and environmental strain can lead to the onset of rheumatoid arthritis.
- *Immunological*—the presence of rheumatoid factor in the serum and synovial fluid of affected patients suggest immunological etiology. Plasma cells and lymphocytes are also present on histological examination.

Clinical Features

General

- *Age and sex distribution*—it more commonly occurs in temperate climate and has its highest incidence in women from 20 to 50 years of age.
- *Sites*—in typical cases, small joints of fingers and toes are the first to be affected. Swelling of the proximal but not the distal, interphalangeal joints give the finger as like spindle appearance and swelling of the metatarsophalangeal joints results in broadening of feet.
- *Symptoms*—symptoms include bilateral stiffness, crepitus, tenderness and swelling over the joint. Fever, malaise, fatigue, weight loss, pain and stiffness in the limb are also evident.
- *Polyarthritis*—polyarthritis develops subsequently, large and weight bearing joints are frequently affected.
- *Subcutaneous or rheumatoid nodules*—there is formation of subcutaneous nodules on the pressure points, sites of friction and various vascular lesions, both necrotizing and obliterative types. Severe deformities of extremity can occur as a result of joint collapse, tendon rupture and muscle involvement.
- *Signs*—the joint may become red, swollen and warm to touch. Muscle atrophy around the joint is common.
- *Hands*—in hands, it may produce an ulnar drift. Bursitis can also occur.

TMJ involvement

- *Acute case*—in acute cases, there is bilateral stiffness, deep seated pain, tenderness on palpation and swelling over the joint. There is limitation in opening of mouth.
- *Referred pain*—pain on biting is referred to the temporal region, ear and angle of mandible.
- *Chronic cases*—in chronic cases, crepitus is the most frequent finding. Functional disturbances like deviation on opening and inability to perform lateral excursions are common.

- *Anterior open bite*—anterior open bite is present due to bilateral destruction and anteroposterior positioning of the condyle.
- *Ankylosis*—fibrous ankylosis of the joint which may be partial or complex occurs in long term.
- *Complication*—subluxation, secondary arthritis, muscular atrophy, and bird-like face can occur.

Radiological Features

- *Joint space*—the pannus may destroy the disc and due to this, the joint space may reduce slightly or substantially, depending on the severity of the condition and the length of time.
- *Condyle*—there is flattening of the head of the condyle. Erosion of the condyle can be seen (Fig. 25-15). In condyle, hollowing out appearance of the cartilage is seen.
- *Articular eminence*—bone destruction which occurs in articular eminence may be slight or confined to the posterior surface or to the inferior convexity. Most of the eminence may be destroyed in severe long continued cases.



Fig. 25-15: Erosion of condyle seen in rheumatoid arthritis (Courtesy Dr Avinash Kshar).

- *Irregular outline of condyle*—as the disease continues, the condylar outline becomes increasingly irregular and ragged. In most severe cases, condyle may be completely resorbed, resulting in loss of vertical support and anterior open bite.
- *Sharpened pencil or mouth piece of flute appearance*—in advanced stages, erosion of anterior and posterior condylar surface at the attachment of the synovial lining occurs, which may resemble a 'sharpened pencil' or 'mouth piece of flute' (Fig. 25-16).

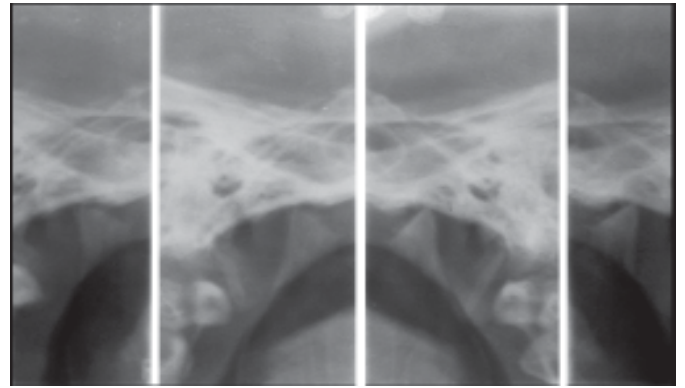


Fig. 25-16: Sharpened pencil appearance seen in rheumatoid arthritis of TMJ.

- *Other features*—subchondral sclerosis and flattening of articular surface may occur with subchondral cyst and osteophytic formation.
- *Radiological staging of rheumatoid arthritis*
 - *Stage I*—periarticular osteoporosis
 - *Stage II*—loss of articular cartilage
 - *Stage III*—erosion (Fig. 25-17)
 - *Stage IV*—subluxation and ankylosis.

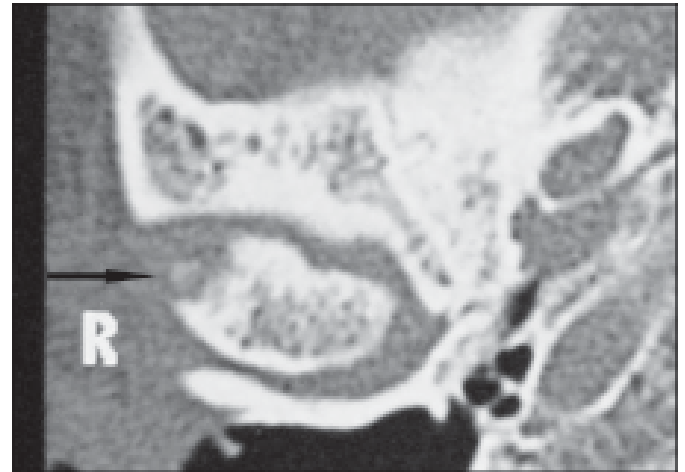


Fig. 25-17: CT scan of erosion of condyle (Courtesy Dr Avinash Kshar)

Diagnosis

- *Clinical diagnosis*—subcutaneous nodule, bilateral involvement and pain on biting. Many joints are involved at the same time.
- *Radiological diagnosis*—sharpened pencil appearance, with flattening of articular eminence.
- *Laboratory diagnosis*—biopsy shows proliferation of synovial lining cells with intense infiltration of lymphocytes, plasma cells and polymorphs. Rice bodies are seen histopathologically. Rose Waller test is positive in 70% of the patients with rheumatoid arthritis. There is elevation of RF factor. Antinuclear antibodies are

detected by indirect immunofluorescence. Analysis of synovial fluid is essential for the immediate diagnosis of joint infection, inflammation and degenerative disease.

Management

- *Supportive treatment*—adequate rest to the joint, soft diet is advocated.
- *Intra-articular corticosteroid injections*—local injection of long acting steroids such as methyl prednisone acetate (20-80 mg for large joint and 4-10 mg for small joint) or triamcinolone hexa-acetomide (10-40 mg for large joint and 2-6 mg for small joint) are given.
- *Non-steroidal anti-inflammatory drugs*—these drugs are inhibitory to prostaglandins. These are used for symptomatic relief. Salicylates (for pain) and anti-inflammatory agents like phenylbutazone, indomethacin, ibuprofen, diclofenac and piroxicam can be used.
- *Immunomodulator*—azathioprine is found to be effective in both, high and low doses.
- *Slow acting anti-rheumatic drugs*—these are the antimalarials like hydroxyl chloroquine sulphate, sulphasalazine (500 mg/day) and methotrexate (D-Penicillamine and parenteral gold).
- *Local treatment*—it is done with heat, diathermy, jaw exercise or a mouth stretcher. Muscle strengthening exercise and hydrotherapy.
- *Medical synovectomy*—synovial obliteration is achieved with osmic acid or variety of radiocolloids, if pain is present even after injection of steroids. Erbium acetate is used for small joints, while yttrium silicate is used for large joints.
- *Surgical synovectomy*—it accounts for removal of synovial membrane which is responsible for enzymatic destruction of cartilage.

Juvenile Rheumatoid Arthritis

It is also called as 'Still's disease' and 'Juvenile polyarthritis'. It is defined as a chronic synovitis with or without extra-articular manifestations, but it is accompanied by more systemic features than for adults.

It occurs in children and has its peak between 1 to 3 years. Initial bilateral polyarthritis of both small and large joints including the cervical spine occurs. There is splenomegaly, lymphadenopathy, leukocytosis, pyrexia and rash.

Patient complaint of neck pain, limited range of movement and restricted opening of the mouth. One or both TMJ are involved. It may cause interference with normal condylar growth, leading to micrognathia. This is seen in at least 20% of cases. Persistence leads to deterioration of jaw movement.

Psoriatic Arthritis

It is a chronic disease of unknown etiology characterized by skin lesions and sometimes joint involvement.

Etiopathogenesis

- *Hereditary*—the exact cause is unknown. In some cases, hereditary is the factor, transmitted as a simple dominant trait.
- *Precipitating factors*—in some cases, the precipitating factors include infection by various microorganisms, metabolic disturbances, endocrine dysfunction, neurogenic factors and trauma.
- *HLA factors*—there is evidence that disease is associated with human major histocompatibility (HLA) antigen complex, implying that either the HLA antigen itself or a linked gene is directly involved in the disease process.

Clinical Features

General

- *Location*—skin lesions are found on the trunk, arms, face and scalp.
- *Appearance*—skin lesions exhibit broad irregular papules or plaques, which are dull red to brownish in color and are usually covered with a layer of fine silvery scales.
- *Auspitz's sign*—when scraped, they leave behind small bleeding points: this is called as Auspitz's sign.
- *Exacerbation*—exacerbation occurs after exposure to ultra-violet light.

TMJ involvement

- *Symptoms*—preauricular pain, which is usually unilateral. There is difficulty in opening the mouth.
- *Signs*—TMJ is tender. Crepitus, deviation towards the affected side and in small proportion of cases, deformities are seen.

Radiological Features

- *Articular surface*—generalized appearance of irregularity of condylar articular surface.
- *Osteoporosis*—generalized osteoporosis can occur.
- *Joint spaces*—proliferative changes with diminution of joint space.

Diagnosis

- *Clinical diagnosis*—positive Auspitz's sign with tender TMJ will give clue to the diagnosis.
- *Radiological diagnosis*—generalized irregularity of the bone is present.

Management

- *Drugs*—Systemic drugs like salicylates should be given.

- **Appliance**—screw wedge appliance should be made and is given to the patient.
- **Steroids**—for skin lesion local application steroids.
- **Others**—shortwave diathermy, massage exercise, screw wedge appliance.

Infective Arthritis

It is also called as 'septic arthritis'. It may be acute or chronic.

Etiology

- **Microorganisms**—it is caused by direct spread of organisms like staphylococci, streptococci, pneumococci and gonococci, from an infected mastoid process, tympanic cavity or via blood.
- **Trauma**—it may also be caused by trauma directly to the joint or infection from a maxillary molar and parotid gland.
- **Osteomyelitis and middle ear infection**—it is acute in nature. It can be caused by osteomyelitis and suppurative middle ear infection.
- **Brucellosis**—brucellosis can also cause infective arthritis usually of chronic type but acute infection can occur.

Pathogenesis

- **Dilatation of blood vessels of synovial membrane**—early dilatation of blood vessels of the synovial membrane with serous exudate is accompanied by the usual stages of inflammation. This is followed by suppuration, with formation of pus and ulceration of the synovial membrane.
- **Erosion of articular disc**—the articular disc is eroded and partly destroyed. The cartilage of the head of the condyle and the fossa are similarly affected.

Clinical Features

- **Age and sex distribution**—it usually occurs in young children with no sex predilection.
- **Location**—it is always unilateral. In some cases of chronic variety bilateral involvement can occur.
- **Symptoms**—there is severe pain on jaw movement, with an inability to place the teeth in occlusion, due to presence of infection in the joint.
- **Signs**—redness and swelling over the joint. In some cases, swelling may be fluctuant and extend beyond the region of the joint.
- **Lymph nodes**—tender cervical lymph nodes on the side of infection. This helps to distinguish septic arthritis from other TMJ disorders.
- **Chronic cases**—it may follow an acute infection, but usually it is chronic. In these cases ankylosis of the joint or facial asymmetry (if the growth centers are involved), may occur.

Radiographic Features

Acute

- **Location**—erosion usually seen on anterior and superior aspect of the condyle.
- **Joint space**—in early stages, width of joint space may be increased by inflammatory distension in early infective period. This will cause hazy appearance in radiographs.
- **Articular cortex**—articular cortex of the condyle may become slightly radiolucent. Discontinuities and subtle irregularities of anterior cortical surface may be seen (**Fig. 25-18**).
- **Osteoporosis**—there is considerable osteoporosis of adjacent parts of the condyle and it may extend to whole of the ascending ramus.
- **Sequestrum**—in some cases, there is formation of sequestrum, which if persistent may lead to some added bone destruction.

Chronic

- **Condensing osteitis**—in chronic cases, there may be peripheral condensing osteitis and approximation of the joint surface, as the articular cartilage is eroded.
- **Bone**—there may be frank bone destruction.
- **Condyle**—a small cup shaped excavation on the anterior face of the condyle, having a smooth or irregular base is seen. In rare cases, whole of the condyle may be lost with varying amount of eminence.



Fig. 25-18: Infective arthritis showing destruction of condylar surface on right side (Dr Kshar).

Diagnosis

- **Clinical diagnosis**—severe pain in the joint, with sign of inflammation over the joint will give clue to the diagnosis.

- *Radiological diagnosis*—destructive change in the condyle will give clue to the diagnosis.

Management

- *Rest to joint*—adequate rest to the joint should be given and patient is asked to do limited movements of joint.
- *Liquid diet*—patient should be kept on liquid diet.
- *Antibiotics and analgesics*—appropriate antibiotics and analgesics should be given to patient.
- *Surgical*—if there is suppuration, incision and drainage should be carried out.

Ankylosing Spondylitis

It is also called as 'Marie-Strumpell disease' and 'rheumatoid spondylitis'. It is a chronic inflammatory connective tissue disease that affects the axial skeleton and central joints including the TMJ.

Clinical Features

- *Age and sex distribution*—it is more common seen in young adults and is more common in men than in women.
- *Site*—it primarily involve spine joint. This will result in forward and stopped posture.
- *Signs*—joint stiffness results from immobility (during sleep) and is typically relieved by heat and exercise.

Radiographic Features

- *Flattening*—there is flattening of the articular surface of the TMJ (Fig. 25-19).
- *Osteophytes*—osteophytic formation is common.
- *Erosion*—in some cases, erosion of condylar head is seen.



Fig. 25-19: Coronal section showing flattening of articular surface (Courtesy Dr Avinash Kshar and Dr Umarji).

Diagnosis

- *Clinical diagnosis*—joint stiffness associated with forward and stooped posture of the spine joint.
- *Radiological diagnosis*—flattening of joint surface.

Management

- *Analgesics*—NSAID should be given to the patient to relieve pain and inflammation.

Traumatic Disorders of TMJ

Condylar Fracture

Condyle is displaced medially, inferiorly and anteriorly due to pull from lateral pterygoid muscle. A blow on the point of chin is the usual cause for bilateral fracture of the condyle.

Classification

Intracapsular Fracture

- *Articular surface fracture*—fractures involving articular surface.
- *High condylar fracture*—fractures above or through the anatomical neck, which may be termed as high condylar fracture.
- *Injury to capsule, meniscus and ligament*—fractures associated with injury to the capsule, ligament and meniscus.

Extracapsular Fracture

- *High condylar fracture*—the fracture runs from the lowest point of the curvature of the sigmoid notch, obliquely downwards and backwards below the surgical neck of the condyle, to the posterior aspect of the upper part of the ramus.
- *Low or subcondylar fracture*—the fracture is influenced in its site and its direction by the insertion of part of masseter muscle and TMJ ligament, the fracture taking the line of least resistance. Such a fracture may be termed as low or subcondylar fracture.
- *Combined fracture*—very nearly the condylar head may be split in the anteroposterior or sagittal plane; the split extending through the neck and producing a combined intra- and extracapsular fracture.

Etiology

- *Indirect violence/fight*—the ultimate position of the fractured fragment will depend on direction and degree of violence, precise point of application, presence or absence of teeth or denture and the extent of occlusion which exists at the moment of impact.

- *Parade ground fracture*—if the blow is sustained centrally on the chin then there is a possibility of bilateral fracture of the condyle. It will protect the vital organs in the middle cranial fossa as the thin neck of the condyle will fracture first. This commonly occurs when a soldier faints on a parade ground and hence named as ‘parade ground fracture.’
- *If the teeth are in occlusion*—there is no displacement of the condyle. If the teeth are not in occlusion then displacement of the condyle is definite.
- *Lateral force*—if the force of violence is received laterally, then the fracture occurs at opposite condyle.

Clinical Classification

- *No displacement*—crack/fracture is present without tearing of periosteum or of ligament and without significant alteration in the normal relationship of the condylar head to glenoid fossa or neck of the condyle to the ramus.
- *Deviation*—angulations exist between the condylar neck and ramus.
- *Displacement*—overlapping occurs between the condylar processes and the surgical fragment lies lateral to ramus.
- *Comminuted fracture*—may occasionally be encountered. They are usually associated with a fracture of the zygomatic arch and coronoid process, due to severe road accidents and gun shot wounds.
- *Greenstick fracture*—it is seen in cases of children. If the blow is not severe, this type of fracture occurs with medial and anterior angulations, which tend to correct itself spontaneously with the natural growth.
- *Dislocation*—fractured dislocation of the condylar head occurs due to pull of the muscle.

Importance of Muscle Pull

- *Detachment of disc*—the TMJ capsule is strengthened on the lateral aspect by the temporomandibular ligament, whereas medially, it is comparatively weak. Therefore when a fracture with dislocation occurs (particularly if the line of force is parallel to the slope of the articular eminence and condylar head is positioned anterior to the eminence), due to the pull of external pterygoid, detachment of disc may occur.
- *Dislocation of condyle in upwards direction*—in a closed mouth, muscle pull will dislocate the condyle upwards and posteriorly on the fractured site. Due to fracture, the force of external pterygoid muscle will be ineffective whereas masseter and internal pterygoid muscle will exert combined upward lift which results in backward movement of the fractured end of the ramus on the uninjured disc.

- *Rotation of condyle on unaffected side*—on the unfractured side, there will be rotation of condyle about a vertical axis. The net result is deviation of the mandible to the injured side. During opening, the external pterygoid of the uninjured side will contract but external pterygoid of the injured side will be ineffective as a result there will be more deviation of the mandible to the injured side.

Clinical Features

General

- *Incidence*—condylar fracture accounts for 18% of mandibular jaw fractures.
- *Location*—unilateral fracture of the condyle is more common as compared with bilateral but both occur with or without injury. Fracture is more often associated with dislocation if it is bilateral.
- *Associated fracture*—most of the fractures of condyle are associated with fracture of the opposite side of the mandibular body.
- *Complication*—complication like arthrosis, arthritis, ankylosis and open bite deformity will occur.

Unilateral fractures (Fig. 25-20)

- *Symptoms*—pain on the affected side; it is increased when movement is attempted. Patient complains that his teeth do not occlude in the normal fashion. Patient is also not able to bring the jaw forward.



Fig. 25-20: Transorbital view of the condylar fracture showing medial displacement.

- *Paresthesia of lower lip*—paresthesia of lower lip can occur in some cases. It occurs as hemorrhage from the condylar region, tracks across the base of the skull and exerts pressure on the mandibular division of the trigeminal nerves as it emerges from the foramen ovale.
- *Signs*—small localized swelling over the injured TMJ. There is tenderness over the condylar area of the affected TMJ. There is also occasional crepitus and deviation of the mandible to the affected side.
- *Ear*—there is bleeding from the ear on the affected side which results from laceration of the anterior wall of the external auditory meatus, caused by violent movement of condylar head against the skin in this region.
- *Ecchymosis*—hematoma surrounding the fractured condyle may track downwards and backwards below the external auditory canal. It gives rise to ecchymosis below the external auditory meatus.
- *Limitation of jaw movement*—there is painful limitation of lateral excursion towards the normal side.
- *Gagging*—there is gagging of the ipsilateral molar teeth.

Bilateral Fractures (Fig. 25-21)

- *Gagging on molar teeth*—all the above signs plus variable degree of gagging of the occlusion on the molar teeth.
- *Restricted mandibular movement*—overall mandibular movements are more restricted, than in unilateral fracture.
- *Forward displacement of mandible*—mandible is displaced forwards in case of bilateral fracture.

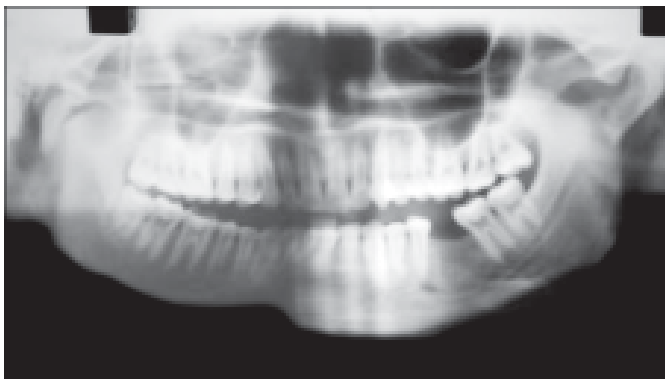


Fig. 25-21: Bilateral condylar fracture seen on OPG of the patient.

Fracture Dislocation

- *Absence of condyle*—all the above findings plus a definite absence of condyle in the glenoid fossa (Fig. 25-22).
- *Unilateral dislocation*—if the condylar head is dislocated medially and all edema has subsided due to passage of time, it may be possible to observe a characteristic 'hollow' over the region of the condylar head.
- *Bilateral dislocation*—if there is a bilateral dislocation, then there is an anterior open bite present.
- *Central dislocation*—it is a rare type of injury which is usually severe and forces the condyle into the cranial cavity through the floor of the articular fossa. It is termed as central dislocation of the TMJ.

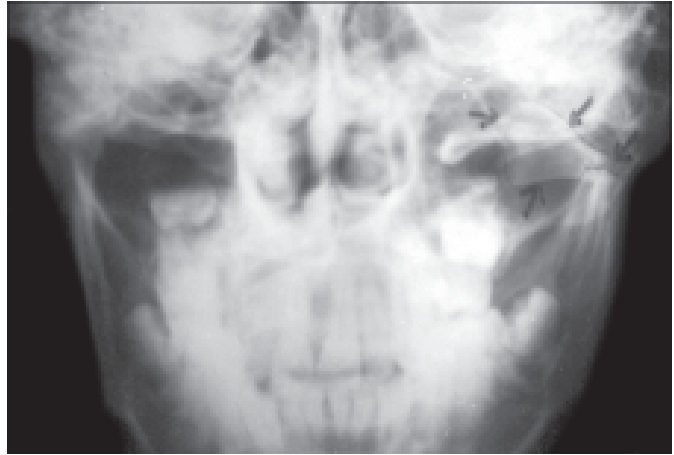


Fig. 25-22: Fracture dislocation showing absence of condyle in the glenoid fossa (Courtesy Dr Datarkar).

Radiographic Features

- *View taken*—displaced condylar fracture is well demonstrated on AP and lateral projection. Non-displaced fracture is well seen on AP view (Fig. 25-23).
- *Increased width of fracture condyle*—when fracture occurs, contraction of the lateral pterygoid muscle rotates the transverse axis of the condylar head, so that the medial end moves anteriorly, thus increasing the apparent width of the fractured condyle.
- *Complete fracture dislocation*—in complete fracture dislocation, the condylar head may be inclined medially at an approximate 45° angle to the vertical axis of the ramus.
- *Anterior displacement of the condyle*—if the condylar head has been displaced anteriorly and turned at 90° angle to the vertical axis of the ramus as viewed from the lateral aspect, then only the articular surface of the condylar head will be seen; this will appear as a narrow radiopaque bar situated in the infratemporal fossa.
- *Condyle split*—the condyle may split with little or no displacement of the fragments, or some portion may be separated from head of the bone. Rarely, the whole of the articular portion is crushed and flattened.
- *CT scan*—in difficult cases, CT scan has been demonstrated to show changes in the relationship of the condyle to the mandibular fossa more precisely than the conventional radiographic examination (Fig. 25-24). It is also claimed that it can demonstrate fine bony alterations at the fractured site.
- *Fracture line*—the fracture line is often transverse but usually oblique, starting in the base of the mandibular

notch and passing slightly or even markedly downwards. In absence of any displacement, it is difficult to visualize such fractures. In the lateral projection, there is often no evidence of any fracture line; but when the posterior margin of the ramus is followed, a sudden step is seen. In some cases there is displacement of the adjacent margins of the fragments, so that the inferior borders of the condylar fragment are superimposed over the adjacent ramus.

- *Articular surface*—the articular surface of the condyle is usually rotated inwards with fracture dislocation.



Fig. 25-23: Subcondylar fracture seen on left side—AP view.

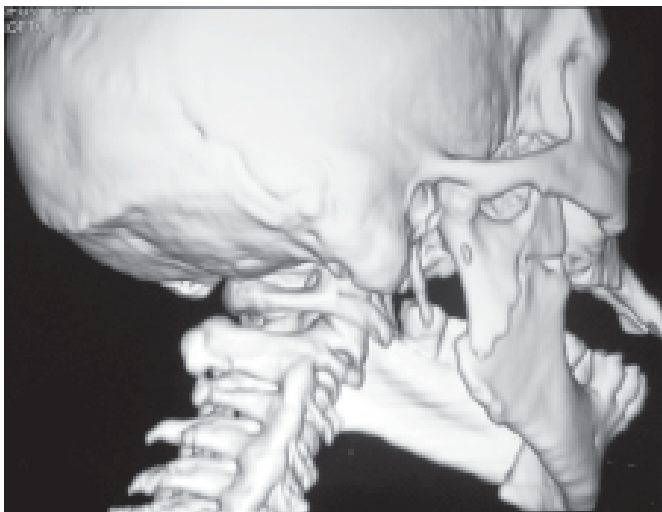


Fig. 25-24: 3D CT scan of patient showing fracture of condyle.

Management

- A *simple crack fracture* without displacement will not require fixation. Patient should be kept under observation and heavy masticatory forces are to be avoided.
- A *fracture with slight displacement* reduction and immobilization of the mandible for a period of 4 weeks.
- *Unilateral fractures with dislocation*—mandible should be immobilized in normal occlusion for ten days then the fixation is released and movement is encouraged. Patients are kept under observation.
- *Bilateral fractures with dislocation*—arch bar is fitted and reduction is achieved by traction in the incisor area. The mandible is immobilized in normal occlusion for 4 weeks.
- *Gunning splints*—for *edentulous patients* gunning splints should be used.
- *Early treatment*—to avoid fibrous ankylosis early movement of TMJ are to be achieved.
- *Other treatment* includes relief of acute symptoms, restoration to proper anatomic relation, intermaxillary fixation for restoration of occlusion and early mobilization to minimize scarring.

Ankylosis

It is also called as '*stiff joint*'. Ankylosis, a Greek word means fusion of body part. It is an abnormal immobility and consolidation of the joint.

Types

- *True (intra-articular)*—it is any condition that produces fibrous or bony adhesion between the articular surfaces of the TMJ.
- *False (extra-articular)*—it is the one which results from pathologic conditions outside the joint, that result in limited mandibular mobility.
- *Bony (true)*—if bone is present between the articulating surfaces and prevents movements, it is called as bony ankylosis.
- *Fibrous (false)*—if the medium which prevent, the movement is fibrous, it is called as fibrous ankylosis.
- *Partial*—if there is incomplete union between the articulating surfaces, it is called as partial ankylosis.
- *Complete*—if there is complete union between the articulating surfaces, it is called as complete ankylosis.

Etiology

False

- *Myogenic*—the most common problem associated with muscle origin is fibrosis, which may result from chronic infection of the elevator muscles of mastication. Myositis ossificans can also produce limitation of opening.

- *Neurogenic*—they include epilepsy, brain tumor, bulbar paralysis and cerebrovascular accidents.
- *Psychogenic*—here, the affected persons exhibit no pain, but patient cannot get the jaws separated also called as hysterical trismus and is apparently produced due to fright.
- *Bone impingement*—the most common is coronoid impingement. Malformation of coronoid such as exostosis or elongation can cause the mandible to impinge on the posterior aspect of the zygoma, when opening is attempted.
- *Trauma*—the formation of fibrous adhesions or cicatricial bands of scar tissues usually occurs after a traumatic accident or burns.
- *Neoplastic disease* like osteochondroma.

True

- *Congenital*—abnormal intrauterine development, birth injuries and congenital syphilis.
- *Trauma*—trauma to the chin forcing the condyle against the glenoid fossa, particularly with bleeding in the joint. Malunion of condylar fractures. Injuries are associated with fracture of the malar-zygomatic compound. In the case of injury to the joint, there is hemorrhage within and outside the joint capsule. Injury occurs during the period of active bone formation. There is immobility of the jaw due to pain or any treatment and ankylosis take place.
- *Inflammatory*
 - Primary inflammation of the joint.
 - Inflammation of the joint secondary to a local inflammatory process (otitis media, osteomyelitis, etc).
 - Inflammation of the joint secondary to a bloodstream infection (septicemia, scarlet fever, gonorrhea).
 - Rheumatoid arthritis is the commonest cause of bilateral ankylosis. Gonococcal arthritis can also cause ankylosis of TMJ.
 - Inflammation secondary to radiation therapy.
- *Others*—loss of tissue with scarring and metastatic malignancy.

Clinical Features

General

- *Age*—it is seen primarily in a young age or between 1 to 10 years.
- *Symptoms*—pain and trismus which is directly related to the duration of ankylosis. Patient also complaint of reduced mouth opening (**Fig. 25-25**).
- *Oral problems*—depending upon the duration, there may be poor oral hygiene, carious teeth and periodontal problems malocclusion (**Fig. 25-26**).

Unilateral

- *Incidence*—unilateral ankylosis is more common than bilateral ankylosis.



Fig. 25-25: Reduced mouth opening seen in patient with ankylosis of temporomandibular joint.



Fig. 25-26: Malocclusion seen in ankylosis patient.

- *Symptoms*—mouth opening is impossible, but the patient may be able to produce several millimeter of interincisal opening.
- *Appearance*—asymmetry of the face with fullness on the affected side (**Fig. 25-27**) and relative flattening on the unaffected side.
- *Deviation of face*—in unilateral ankylosis, patient's face is deviated towards the affected side.
- *Shifting of midline*—the chin is retracted on the affected side and slightly bypasses the midline (**Fig. 25-28**). Slight gliding movement towards the affected side.
- *Cross bite*—cross bite is present due to deviation of mandible and shifting of midline.

Bilateral

- *Bird face appearance*—face is symmetrical with micrognathia. There is bird face appearance (**Fig. 25-29**).



Fig. 25-27: Fullness of face seen on affected side (left) due to ankylosis of TMJ.



Fig. 25-28: Shifting of midline on affected side seen in ankylosis patient.



Fig. 25-29: Bilateral ankylosis showing bird face appearance (Courtesy Dr Lambade).

- *Symptoms*—no gliding movement. With bilateral ankylosis, neither protrusive nor lateral movements are possible.
- *An attempt at forced opening*—in bony ankylosis, there is no pain but in case of fibrous ankylosis, there is pain.

- *Muscles of mastication*—due to long standing ankylosis, atrophy or fibrosis of muscles of mastication may result.
- *Congenital ankylosis*—in case of congenital ankylosis, there is difficulty in introducing the nipple into the mouth of newborn infants.
- *Signs*—class II malocclusion (Fig. 25-30), protrusive incisors and anterior open bite. It is due to the patient's attempt at forcing food through the anterior teeth for a period of years. Patient also suffer from micrognathia (Fig. 25-31).



Fig. 25-30: Protrusive incisor seen in bilateral ankylosis.



Fig. 25-31: Micrognathia seen in patient with bilateral ankylosis.

Radiographic Features

Fibrous ankylosis

- *Appearance*—in some cases, there is transverse or oblique dark line, irregular in outline, crossing the mass of dense bone (Fig. 25-32). When a dark line is present, the possibility of fibrous ankylosis is more.

- *Ramus and body of mandible*—the ramus is vertical and the angle is reduced in size. The body of the jaw is short
- *Condyle*—the condyle may retain its normal shape, but it can be replaced by shapeless mass of bone, which finds attachment to the base of skull above and to the base of neck of condyle below.

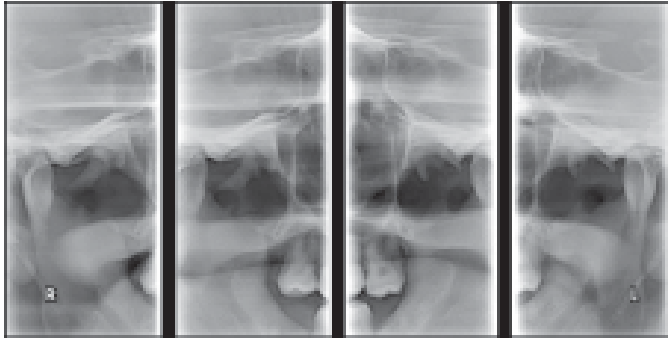


Fig. 25-32: Fibrous ankylosis seen in patient showing dark line crossing the bone.

Bony ankylosis

- *Joint space*—joint space is completely or partially obliterated with dense sclerotic bone (**Fig. 25-33**). Sometimes, large mass of new bone may be seen, radiographically obscuring the condyle and joint space.



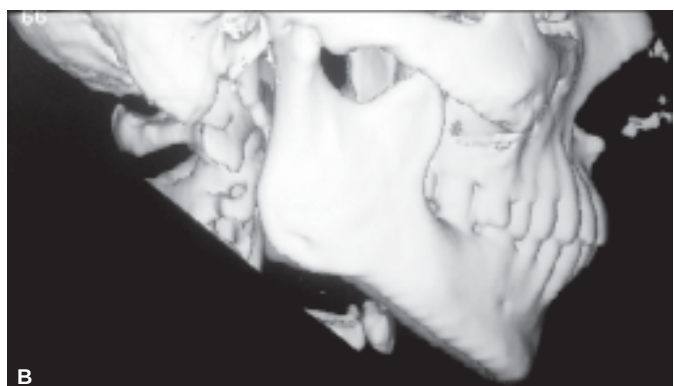
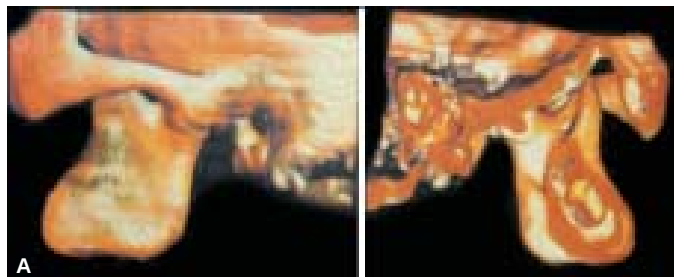
Fig. 25-33: Complete obliteration of joint spaces. seen in bilateral bony ankylosis.

- *Antegonial notch*—prominent antegonial notch on the affected side of mandible (**Fig. 25-34**), along with inferior arching of the mandibular body (secondary to isotonic contraction of the depressor muscles).
- *Condyle*—bone may form around the neck of the condyle and becomes continuous with the base of the skull. Considerable destruction of bone may precede bony ankylosis. The greater part of the condyle may have been destroyed so that the sigmoid or mandibular notch is approximated to the base of the skull. The neck of the condyle, if not completely hidden by the mass of new bone, appears to be shortened; so that the mandibular notch is nearer the zygomatic process than normal. No translation of condyle head during opening.



Fig. 25-34: Prominent antegonial notch seen on right side of patient in unilateral ankylosis

- *Dark linear shadow*—in some cases, there is horizontal, slightly irregular, dark linear shadow in the middle of the new bone, which represents the cartilage and meniscus.
- *Coronoid process*—there may be deepening of the notch which has escaped involvement in the new bone formation and the appearance may be accentuated by the elongation of the coronoid process, which at the same time is narrow and slender.
- *Computed tomography*—this is nowadays valuable aid in determining the extent of ankylosis (**Figs 25-35A and B**).



Figs 25-35A and B: 3D CT of ankylosis of temporomandibular joint.

Diagnosis

- *Clinical diagnosis*—in bilateral case, bird face appearance or anterior open bite is present. In unilateral cases, deviation of mandible, shifting of midline is present.

- *Radiological diagnosis*—absence of joint space with prominent antegonial notch will diagnose this condition.
- *Laboratory diagnosis*—atrophic or destructive changes in the cartilaginous component of the joint with loss of meniscus are a constant finding.

Management

- *Brisement force*—forceful opening of the jaws under general anesthesia.
- *Condylectomy*—condyle is excised.
- *Gap arthroplasty*—it is performed in the mandibular neck. Two parallel lines are cut, beginning in depth of the sigmoid notch and carried downwards at an angle of 45 degree at the posterior border of the ascending ramus.

Dislocation

It results when condyle is forcefully displaced anteriorly (by failure of muscular co-ordination) out of the articular fossa but remains within the capsule of joint. The direction of condyle is almost always anterior, beyond the articular eminence.

Classification

- *Anterior*—it is more common than other two.
- *Posterior*—it is rare and results from the severe injuries which are sustained at the point of the chin or at the inferior border of the body of the mandible.
- *Central*—same as the posterior dislocation
- *Medial or lateral*—it occurs due to injury to the neck of condyle with its fracture. It is very rare.

Etiology

- *Extrinsic force*—it can occur under G.A. following extraction of teeth or the use of mouth gag, when muscle relaxant is used.
- *Intrinsic*—these are related to attempts of excessive opening of the mouth during eating or yawning and occasionally singing. These types of dislocation are sometimes called as *spontaneous dislocations*.
- *Occlusal disharmony*—in cases of *recurrent dislocation*, the etiologic factor is different, like occlusal disharmony producing muscle spasm.
- *Phenothiazine*—occasionally, action of phenothiazine group of drugs produces spontaneous dislocation of the mandible or spasm of the facial musculature.
- *Other causes* like epileptic seizure, yawning, taking too large bite of an apple and rarely, followed sneezing.

Clinical Features

Anterior dislocation

- *Site*—anterior dislocation may be unilateral or bilateral. In acute dislocation, there is a history of injury, gagging of molar teeth and anterior open bite.
- *Symptoms*—patient has great difficulty in swallowing and saliva drools over the chin. Pain in the region of temporal fossa and there is a depression where the condylar head is normally situated.
- *Signs*—the mandible is postured forward and movement is extremely limited. The condyle becomes locked anterior to the articular eminence and is prevented from sliding back by muscular spasm. When unilateral dislocation occurs, the teeth will be gagged posteriorly on the side of dislocation and the chin will be deviated towards the normal side.

Central dislocation

- *Cause*—the head of the condyle is forced through the floor of the articular fossa, into the floor of middle cranial fossa. In this case, there may be damage to the articular surface of the condyle and fracture of the neck.
- *Neurological signs*—neurological signs like the cerebral contusion, facial nerve paralysis, deafness and bleeding from the ear may also be present.

Posterior dislocation

- *Cause*—it results from severe injury, which forces the mandible backwards, so that the condyle penetrates the tympanic plate of the temporal bone. When the upward and backward thrust is applied to the chin, posterior dislocation occurs.
- *Age*—it is more common in elderly people as they have increased gonial angle, which leads to more posterior displacement of the condyle than in the case of an individual having a smaller angle.
- *Symptoms*—there may be tenderness over the affected TMJ, limited mouth opening, anterior open bite and laterognathism.

Radiographic Features

- *Anterior dislocation*—it is difficult to detect radiographically, as there is a wide variation in anterior condylar movements. Condyle is locked in front of the articular disc. Condyle is high on the anterior surface of the eminence and is well up in the infratemporal fossa and stayed there when the patient have closed their mouth.
- *Central dislocation*—radiologically, the anatomy of the articulation is seen to be altered, so that neck of condyle occupies a too high position in relation to the articular fossa.

- *Posterior dislocation*—radiologically, the anterior portion of the articular fossa is empty and the condyle is seen too far posteriorly. The neck of the condyle may appear short, if the head of condyle maintain its abnormal position. In such a case, the condyle may be seen partly covered by the more radiolucent external auditory canal. In cases when there is posterior dislocation into the tympanic plate, but has been subsequently reduced, it may be possible to identify the damage to the plate in good quality radiograph.

Diagnosis

- *Clinical diagnosis*—anterior open bite with gagging of molar teeth will aid in diagnosis.
- *Radiological diagnosis*—empty articular fossa space will diagnose this condition.

Management

- *Reduction of dislocation*—it can be done by the following
 - *Manipulation without anesthesia*—some type of sedation should be given to the patient before manipulation. Patient should be sitting on the chair with the operator in front. The thumbs covered with gloves should be pressed down over the buccal shaft area of the mandible, near the lower molar teeth and at the same time elevating the chin with fingers and pushing the entire mandible posteriorly and then upwards.
 - *With local anesthesia*—this treatment is based on the principle, that the dislocation is maintained by muscle spasm secondary to painful stimuli, arising from the capsule. The treatment consists of injection of local anesthesia (lignocaine hydrochloride) into the joint. After approximately one minute spontaneous reduction generally occurs without the necessity of manual reduction. Even in bilateral dislocation, unilateral injection is adequate. This is due to the elimination of neural reflex which extends over the muscle bilaterally and patient can close the mouth to normal position.
 - *Under general anesthesia, with the aid of muscle relaxant*—if the above mentioned method fails and the patient is unduly apprehensive then this method is used. Postoperatively, the jaws should be immobilized for 10 to 14 days, this help the inflammation and edema to subside.

Subluxation (Hypermobility)

It is the unilateral or bilateral positioning of the condyle anterior to the articular eminence, with repositioning to normal accomplished physiologic activity. It is self reducing

incomplete dislocation, which generally follows stretching of the capsule and ligaments.

Etiology

- *Long continuous opening of mouth* in the dental chair or singing, yawning or sleeping on one arm in bed.
- *Oral surgical procedures* excessive movement apparently causes stretching of the joint ligament or rupture of the external pterygoid attachment to the meniscus.
- It may follow the chronic degenerative changes of *osteoarthritis*.
- It may be a manifestation of an underlying *psychiatric problem*, which may be manifested by minor trauma.
- Use of *phenothiazine* derivatives by surgeons may also be responsible (due to hypersensitivity to the drug), it returns to normal after anti-histaminic therapy.

Clinical Features

- *Symptoms*—cracking noise, temporary locking of the condyle and immobilization of the jaw. Patient describes weakness of the joint while yawning. Pain is associated with last few millimeters of mouth opening.
- *Signs*—the condyle may get locked when the mouth is opened widely and upon closing, it will return with a jumping motion, accompanied by a sound caused by movement of the condyle over the articular eminence. On palpation 'click' on opening and sliding of condyle over the articular eminence, are common.

Radiological Features

Excessive excursion of the condyle from rest position to the position when jaw is opened wide (**Fig. 25-36**).

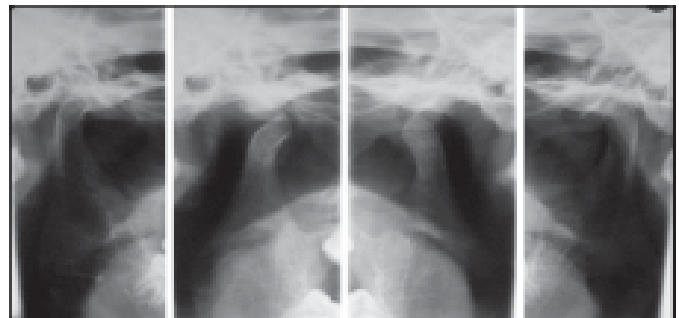


Fig. 25-36: Subluxation of TMJ observed on panoramic radiograph (Courtesy Dr Tapasya Karamore).

Diagnosis

- *Clinical diagnosis*—cracking noise will temporary locking of condyle will give clue to diagnosis.
- *Radiological diagnosis*—excessive excursion will be seen radiologically.

Management

- *Conservative method*—here, the objective is to achieve shrinkage of the capsule by a sclerosing agent, which will cause fibrosis of the capsule. It will limit the condylar excursion without deleterious effects on motion and cartilaginous part of the articulating surface of the condyle. The solution used is 5% sodium psylliate or 5% intracaine in oil base. A mixture of equal parts of 0.5% of eucupine in oil and 5% aqueous solution of sodium psylliate has an additional effect. It may be necessary to repeat the injection every 2 to 3 weeks, till fibrosis occurs.
- *Surgery*—insertion of bone graft is done. The joint is exposed; a vertical incision is made in the outer side of the capsule. The edges are overlapped and then sutured so as to tighten the capsule in the anteroposterior plane. After 7 days of intermaxillary fixation, clicking and subluxation will be relieved. Shortening of the temporalis tendon also relieves subluxation.

Internal Derangement or Disc Displacement

Here, the disc is abnormally located in relation to other components of the joint. It is usually displaced antero-medially. In some rare cases, disc may be displaced posteriorly. Most of the cases of disc displacement occur due to microtrauma from bruxism or clenching.

It can be defined as a malrelationship of the meniscus to the condylar head and articular eminence, where an alteration of its attachment allows the meniscus to assume an abnormal position.

Classification

- *Anterior displacement with reduction (Fig. 25-37)*—In it, as the condyle moves forward, it snaps under the posterior edge of disc producing an audible click and on closing the condyle may again snap under the posterior edge of the disc producing another click.
- *Anterior displacement without reduction*—There is no reduction of condyle in the mandibular fossa in closed position.
- *Posterior disc displacement*—condyle slip over anterior rim of disc during opening.
- *Disc displacement with intermittent locking.*
- *Disc displacement with perforation.*

Etiology

- *Articular surface remodelling*—it occurs due to combination of articular surface remodelling, disc deformation and displacement of either the condyle or the disc from an ideal relationship, which leads to jamming of the disc.

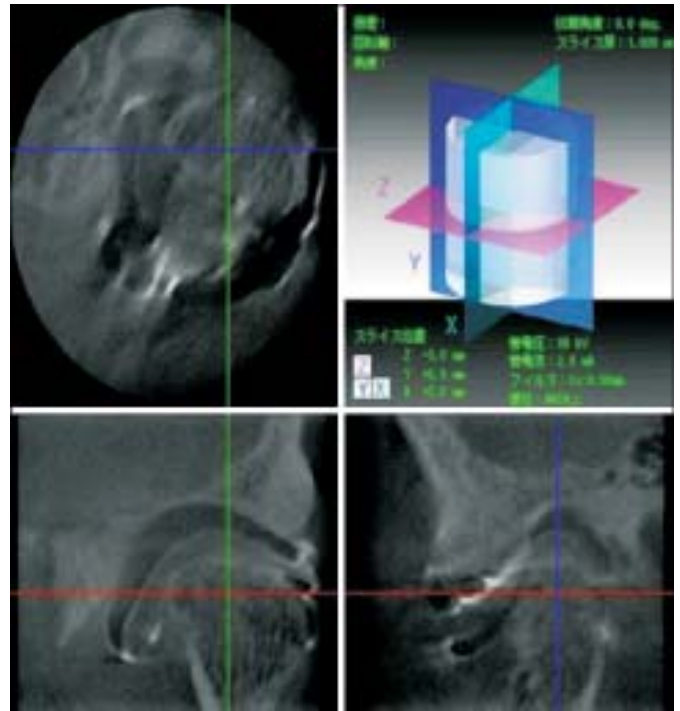


Fig. 25-37: Double contrast CBCT arthrography—it demonstrated anterior displacement of the disc without perforation and dilation of the superior joint space. Also, demonstrates multiple loculi in the anterior region of the joint space (Courtesy Kazuya Honda, Japan).

- *Articular eminence*—very steep articular eminence increases the risk of disc displacement.
- *Disc morphology*—if the morphology of the disc is altered and if the discal ligaments are elongated, there is increased risk of disc displacement.
- *Trauma*—trauma due to malocclusion results in the spasm of the lateral pterygoid and produces a series of changes.

Pathogenesis

- *Early changes*—Increased muscle tone of the lateral pterygoid → it tends to pull the meniscus anteromedially to the condylar head → the meniscus will move anterior to the condylar head → a click or pop will then occur on early opening as the condylar head reseats into the thin central portion of the meniscus → this type of joint noises occur in early stages of joint disease and can be corrected by relieving the muscle spasm.
- *Creation of freely moving disc*—discal ligaments are not elastic and hence once elongated; they generally remain at that length → the elongated discal ligament and the continuous muscle pull due to spasm results in freely moving disc on the articular cartilage.
- *Alteration of joint function*—if the posterior border of the disc becomes thinned and the retrodiscal laminae and lateral discal ligament becomes elongated → the disc can be translated across the articular surface of the

condyle—this translatory movement between the disc and the condyle is abnormal—once the disc is displaced anteromedially, normal joint function is altered.

- *Functional displacement*—at rest, the resistant force provided by the superior discal lamina will act and hence the condylar head will be positioned on the posterior border of the disc (known as functional displacement). Patient has feeling of tightness at this stage. When the person bites, the muscle pull further stretches the disc, causing arthralgia pain.
- *1st click*—in the closed joint position, the condyle articulates on the posterior border the disc. During opening, the condyle moves forward along with the disc. During this opening movement, the disc suddenly moves posteriorly, so that the condyle establishes a normal relationship with the intermediate zone of the disc. This sudden movement between the condyle and the disc results in clicking sound.
- *Reciprocal click*—if the condition persists, then there is elongation of the retrodiscal lamina. At this stage, during closing movements, the condyle moves the intermediate across the posterior border of the disc and just prior to the closed positions a second click is heard. This stage of derangement is called as the reciprocal click.
- *Anterior dislocation with reduction*—the patient can open the mouth to a certain extent at which they have a feel of locking. When the patient moves the jaw away from the painful side, opening is suddenly possible as the jaw suddenly becomes free with a pop. The meniscus is present anterior to the condylar head in the closed position. During mouth opening, the meniscus is pushed in front of the condyle until it mechanically blocks any further excursion. In order to reduce the dislocation, the patient needs to move the condylar head in the direction taken by the meniscus under the influence of the muscle. As the normal relationship is established, full opening is possible. When the mouth is opened, in disc dislocation, the condyle moves onto the posterior border of the disc. This creates jamming or catching sensation during opening. In some instances, the patient can move the mandible medially or laterally and abruptly move the condyle over the intermediate zone of the disc. During jaw movements, the disc returns to the normal position with the condyle.
- *Anterior dislocation without reduction*—in the next stage, due to continuous stretching caused by dislocation, soft tissue degeneration of the joint takes place and the meniscus becomes distorted and hypertrophied. The remaining elasticity of the bilaminar zone is lost and the meniscus remains anteromedially blocking forward the translation of the condylar head. This results in restricted movements and reduction is no longer possible.
- *Perforation of the disc*—due to continuous stretching, the condylar head is positioned on the retrodiscal tissues.

Due to pressure, tissue breakdown begins with inflammation of the tissue, which is called as retrodiscitis. Due to attenuation perforation occurs in the disc, leading to bone-to-bone contact, which ultimately results in osteoarthritis.

- *Disc dislocation*—if the posterior border of the disc becomes thinner and the retrodiscal lamina and collateral ligament become more elongated, the disc can slip anteriorly through the discal space. This is known as disc dislocation, as there is loss of contact between the articular surfaces of the condyle and the disc.

Clinical Features

Anterior disc displacement with reduction

- *Symptoms*—during the disc dislocation, the condyle may articulate on the retrodiscal tissue and may cause pain.
- *Signs*—on physical examination, the joint is tender and reciprocal audible clicking is heard. The jaw is deviated towards the side of the click, till the click occurs and then returns to the midline (**Figs 25-38A and B**).



Figs 25-38A and B: Deviation of jaw on right side in disc displacement with reduction.

Anterior disc displacement without reduction

- Patient who experiences repeated disc dislocations with reduction may further elongate discal ligament and retrodiscal lamina. Often, elasticity of the retrodiscal lamina is lost.
- *Closed lock*—if the condyle moves forward but the disc is not returned to its normal relationship between the condyle and fossa, a condition known as dislocation without reduction occurs. It is also called as closed lock.
- *Symptoms*—there is joint pain, limited opening, previous clicking with intermittent locking and sensation of something in the joint. The joint locks during opening of the mouth.
- *Signs*—there is joint tenderness with deviation towards the affected side. Terminal stretching produces increased joint pain.
- *Mouth opening*—there is limitation of lateral movement in the joint.

Disc displacement with perforation

- *Symptoms*—there is joint pain, clicking with intermittent locking and closed lock. There is also limited opening.
- *Signs*—crepitus and grinding noise. There is also joint tenderness.

Posterior disc displacement

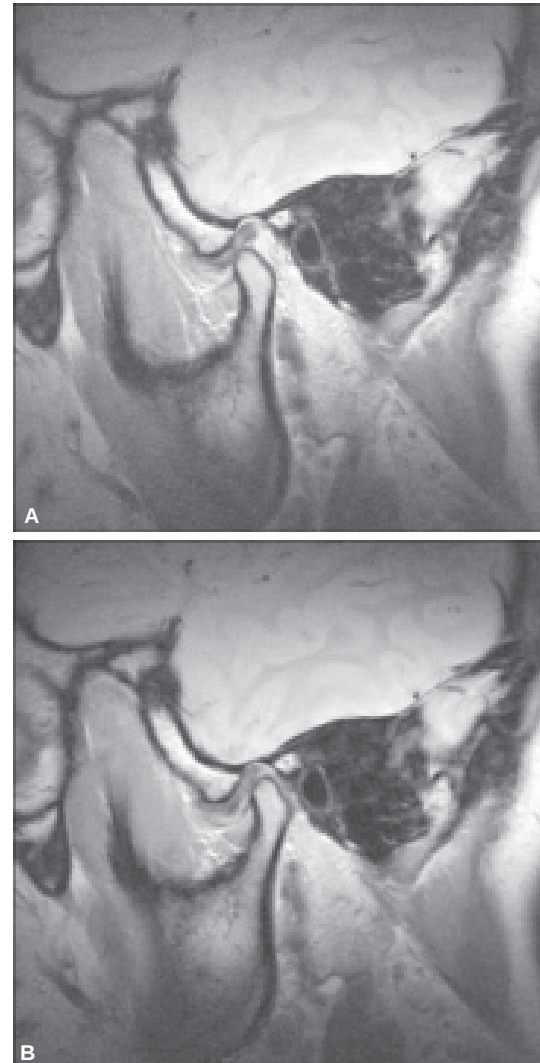
- *Occlusal derangement*—sudden inability to bring upper and lower teeth tooth in maximal occlusion.
- *Pain*—there is pain in the affected joint when teeth are tried to bring in occlusion.
- *Forward displacement*—there is forward displacement of mandible on the affected side.
- *Restricted lateral movement*—there is restricted lateral movement to the affected side.
- *Mouth opening*—there is no restriction of mouth opening.

Radiographic Features

- *Plane radiography*—they are not diagnostic; except in cases of perforation of the disc, where there is evidence of degenerative changes in the joint.
- *Arthrogram*—arthrogram is useful in studying the changes.
- *MRI*—in anterior disc displacement, posterior band of the disc located anterior to the superior portion of the condyle is seen in closed mouth sagittal image. In some cases bone marrow edema can be seen (Figs 25-39 and 25-40).

Diagnosis

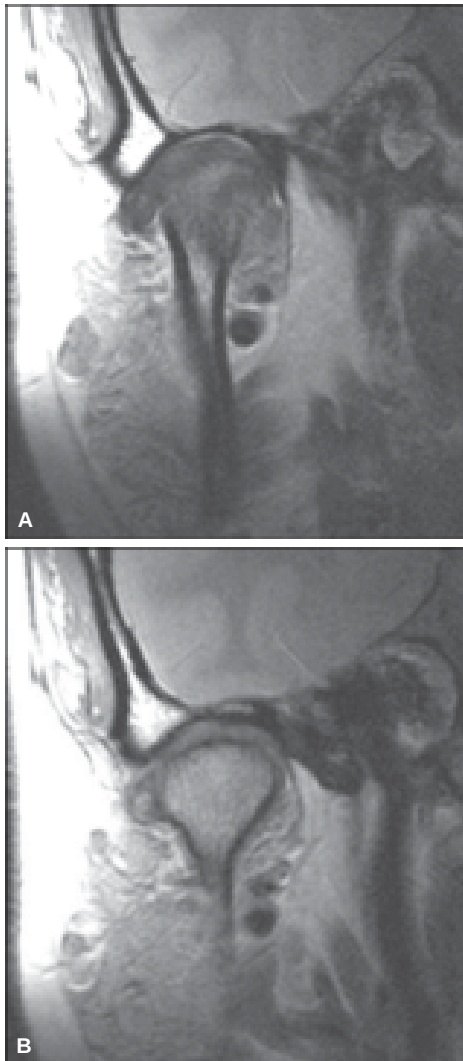
- *Clinical diagnosis*—clicking sound while opening and closing the mandible with pain in TMJ area.
- *Radiological diagnosis*—MRI is useful in the diagnosis of disc displacement.



Figs 25-39A and B: (A) Axial section, closed mouth, anteriorly displaced disc. (B) Axial section, open mouth, anteriorly displaced disc, but normal disc condyle relationship (Courtesy Dr Avinash Kshar and Dr Umarji, Government Dental College and Hospital, Mumbai).

Management

- *Bite plane appliance*—it involves one of the bite plane appliances that position the mandible so that the mouth remains slightly open vertically and is placed anteriorly to maintain the disc in a normal relationship to the condyle. Maxillary and mandibular appliances can be used for maintaining the disc reduction during function. The evaluation and adjustment in the appliance should be made monthly, till the condyle return to normal hinge position. TMJ should be stabilized in the position for two months by maintaining the patient on splints.
- *Occlusal adjustment, restorative and orthodontic treatment*—next phase is of occlusal adjustment, restorative treatment and orthodontics or all the three can be done.
- *Surgical treatment*—it is indicated in those patients who have chronic internal derangement, have failed to respond to non-surgical treatment. It includes



Figs 25-40A and B: Coronal section, closed mouth, laterally displaced disc (Courtesy Dr Avinash Kshar and Dr Umarji, Government Dental College and Hospital, Mumbai).

- **Disc repositioning procedure**—in this, there is removal of tissues from the posterior attachment of the disc with repositioning of disc in its normal position. It is sometimes combined with high intracapsular condylar reduction or an articular eminence re-contouring.
- **Menisectomy**—when it is not possible to repair the disc, interposition material is placed.
- **Postoperative physiotherapy**—postoperative physiotherapy is advocated to establish full range of motion. Bite plane appliance is used for a week followed by occlusal adjustment.

Adhesions

The movements between healthy articular surfaces occur without friction. Changes in the articular surfaces and synovial fluid can drastically change this frictionless system. This sticking of articular surfaces has been called

as *adhesion*. Adhesions can occur in either superior or inferior joint space and can occur with or without disc derangement. Adhesion can begin after prolonged static loading of the joint. The patient wakes up with limited jaw movement. As the patient attempts to move the mandible, a single click is felt (representing freeing of adhesion) and then, the normal range movement is restored. The click only occurs once and cannot be repeated without prolonged period of static loading. Adhesion are best demonstrated on arthroscopy (**Fig. 25-41**).

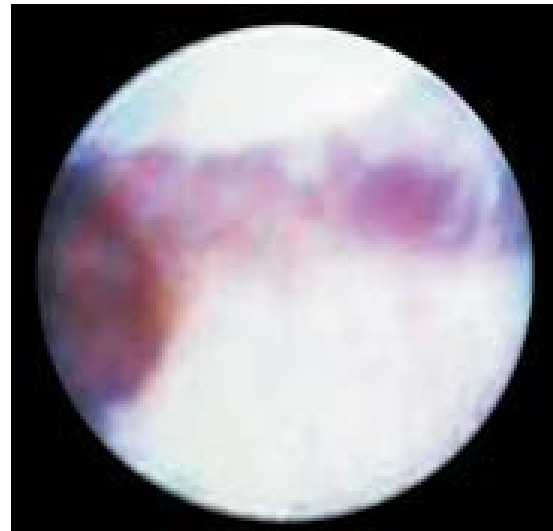


Fig. 25-41: Arthroscopic observation—band-like fibrous adhesion is observed in the posterior articular cavity (Courtesy Kazuya Honda, Japan).

Metabolic Disorders

Gout

It is a chronic metabolic disorder characterized by acute exacerbations of joint pain and swelling associated with an elevated blood uric acid and deposition of crystals of monosodium urate.

Various drugs like thiazide diuretics, operations, trauma, alcohol and rapid weight loss can predispose to this disease.

Clinical Features

- **Age and sex distribution**—seen in middle age with equal sex distribution. It has a hereditary tendency.
- **Sites**—initially, metacarpophalangeal joints are commonly involved. Later foot, ankles, hand, wrist and elbow may be affected.
- **Symptoms**—there is excruciating pain, which may be worsen at night. Sometimes, it is associated with anorexia, fever and general malaise.
- **Progress**—joint returns to normal after few days, with desquamation of the overlying skin.

- *Chronic cases*—as the disease becomes chronic, pain and stiffness persist, with irregular swelling. Tophi are found in cartilage of the ear, nose or eyelids. In half the cases, palms of hands may show white streaks along the creases.
- *TMJ involvement*—sudden excruciating pain in the TMJ, followed by rapidly developing swelling. There is also tenderness of the affected area with limitation of movements. Deviation to the affected side while opening the jaws.

Radiographic Features

- *Punched out lesion*—punched out areas in the carpal bone of the hands. In TMJ also, punched out radiolucency can be seen on the condylar cartilage.
- *Destruction of cartilage*—severe destruction of the cartilage can also be seen in some areas.
- *Calcification in the joint*—there is calcification within the joint spaces which may be subtle or massive.

Diagnosis

- *Clinical diagnosis*—multiple joint involvements. Tenderness on the TMJ area can diagnose the disease.
- *Radiological diagnosis*—punched out lesion with calcification in the joint.
- *Laboratory diagnosis*—articular cartilage reveals white chalky deposits. This deposit is also often found in the synovial and adjacent tissues. The cartilage may be fragmented. Crystalline deposits of uric acid are seen. This is followed by small accumulations (*microtophi*) with surrounding granulomatous foreign body reaction.

Management

- *Diet*—diet should be low in uric acid and fat, i.e. sweetbread, meat, extract peas, beans.
- *Uricosuric agents*—increased elimination of uric acid by uricosuric agents like colchicine 0.5 mg every 2 hourly, to a maximum of 6 mg in 24 hours.

Neoplastic Disorders

Benign Tumors

Benign tumors reported are osteoma and osteochondroma.

Clinical Features

- *Symptoms*—there is stiffness of the joint with facial asymmetry.
- *Signs*—deviation of mandible on the affected side. Disordered occlusion, little movement of the condyle on palpation. There are restricted movements of TMJ.

Radiological Features

- *Appearance*—there is bony mass protruding from the condyle (Figs 25-42 and 25-43).

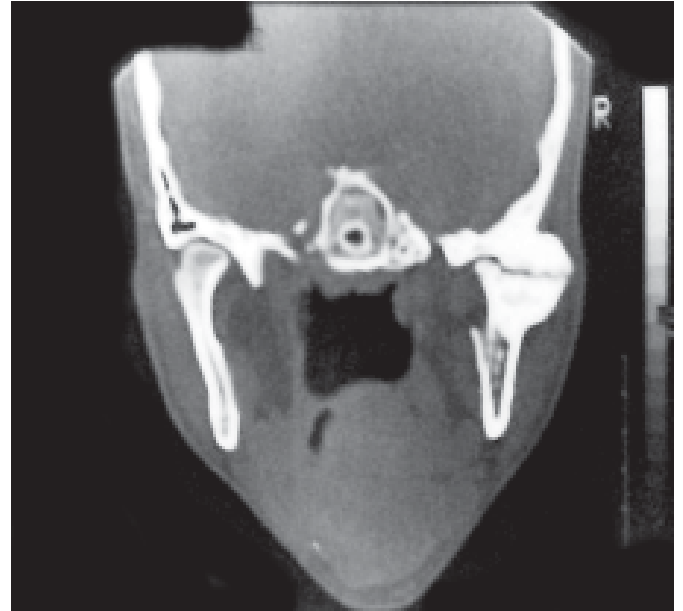


Fig. 25-42: Chondroma of condyle showing radiopaque lesion on right condyle.

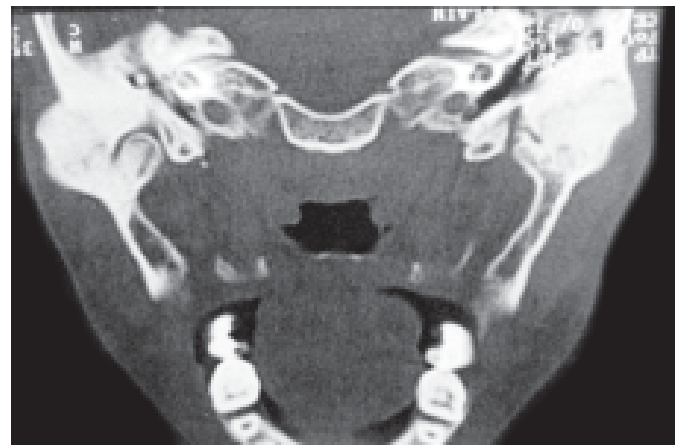


Fig. 25-43: Osteoma of condyle on right side (Courtesy Dr Datarkar).

Management

- *Surgical*—surgical excision of the tumor and orthodontic treatment, to re-establish the occlusion.

Malignant Tumors

Types

- *Intrinsic*—it arises from the bone of the condyle, articular fossa and the joint capsule or the articular disc. It includes chondrosarcoma and synovial sac sarcoma.
- *Extrinsic*—the tumors which extend from outside of TMJ, e.g. neoplasm of the parotid gland.

Clinical Features

- *Age and sex*—age of incidence from 17 months to 68 years with male to female ratio being 1:1.
- *Symptoms*—pain on full opening of the mouth and diminished hearing. Swelling of the TMJ is present. Swelling may be huge to cause facial asymmetry.
- *Signs*—the tumor may be fixed to deep structures. Deviation of the mandible to the unaffected side. There is also malocclusion (Fig. 25-44).



Fig. 25-44: Huge swelling seen on right side of patient due to chondrosarcoma of condyle.

Radiological Features

- *Appearance*—irregular destruction of bone in condylar region is present (Fig. 25-45).

Management

- It can be treated by surgery, chemotherapy, radiotherapy and combination therapy.



Fig. 25-45: Irregular destruction seen in the condylar area due to chondrosarcoma.

Miscellaneous Disorders

Synovial Chondromatosis

It is a benign chronic progressive metaplasia that will not resolve spontaneously. Although it is non-neoplastic, it may resemble a malignant condition histologically. It is also called *chondrometaplasia*. It denotes the condition whereby a cartilaginous focus develops within the synovial membrane of the joint.

Pathogenesis

- *Predisposing factors*—trauma may be a predisposing factor, other factors are malocclusion, occlusal habits, and subluxation or tension sites.
- *Metaplasia of mesenchymal cell*—this is generally believed to occur through metaplasia of mesenchymal cell rests in the underlying connective tissue of the membrane.
- *Formation of metastatic foci*—there is formation of metastatic foci.
- *Detachment of metastatic foci*—foci are detached from the affected membrane and become cartilaginous, and forms mobile bodies within the joint cavity.
- *Calcification of cartilaginous foci*—many of these cartilaginous foci then undergo calcification. These joint bodies acquire a perichondrium, which enables them to grow by proliferation of chondrocytes.

Clinical Features

- *Age and sex*—female to male ratio is 3:1 with greatest incidence at 40-60 years of age.
- *Site*—this affect large joints like knee, elbow, hip and shoulder. This disease is also present in temporomandibular joint.
- *Symptoms*—facial pain, limitation of motion and deviation towards the affected side.
- *Signs*—crepitus, preauricular swelling, enlarged joint with effusion and local tenderness.

Radiographic Features

- *Loose bodies*—this consists of rounded /irregular shaped radiopaque structure in the joint (Fig. 25-46).
- *Joint space*—there is irregularity in the joint space. Joint space is also widened.
- *Condyle*—condylar head is also irregular.

Diagnosis

- *Clinical diagnosis*—it does not show any diagnostic features
- *Radiological diagnosis*—radiopaque loose bodies seen in the joint space.

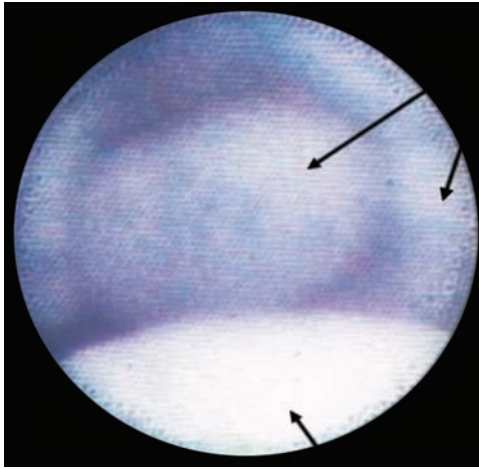


Fig. 25-46: Loose bodies seen as rounded shaped in arthroscopic view (Courtesy Dr Honda).

- *Laboratory diagnosis*—cartilage shows hyperchromatic and binucleated chondrocytes.

Management

- *Surgical*—loose bodies should be removed.
- *Synovectomy*—total synovectomy can be done to prevent recurrence.
- Removal of metaplastic foci and synovectomy are the preferred treatment.

Drug Induced Disorders (Steroids)

Corticosteroids induce mandibular osteoarthritis. The manifestations depend on the dosage, potency and duration of exposure to excessive steroids. Impaired connective tissue function may lead to calcification of the cartilaginous matrix, resulting in osteoarthritis like changes.

Lesions develop initially in the midst of condylar cartilage and then spread to involve the articular cartilage. The effect of steroids on the chondrocyte is inhibitory in nature. The catabolic activity of steroids on the condylar cartilage is due to its inhibitory effect on DNA and RNA synthesis in chondrocytes, with subsequent interference in protein synthesis.

In addition, triamcinolone was found to enhance the synthesis and secretion of parahormone in various experimental animals and thereby could elicit the degenerative changes in the joint tissue.

TMJ Dysfunction Syndrome or Myofascial Pain Dysfunction Syndrome

When muscle spasm develops, dysfunction as well as pain occurs and the condition usually is designated as MPDS. It is initiated as spasm of one or more masticatory muscle.

Etiology

- *Abnormal occlusion*—in case of painful and potentially damaging contact in the occlusion, persons make modifications in the pathway of closure. These corrective movements of the mandible, during closure may elicit muscle strain and spasm, which in turn results in an abnormal pressure on the TMJ.
- *Tooth muscle theory*—occlusal interferences cause an altered proprioceptive feedback, leading to incoordination and spasm of some of the muscles of mastication.
- *Prosthetics problems*—some authors think that interceptive contact, due to faulty complete and partial dentures, over closure, bilateral loss of molar teeth, and increased vertical dimension in partial and complete denture may lead to TMJ dysfunction. There is a change in the myotactic stretch reflex in all above mentioned conditions, which support the view that the TMJ dysfunction can result from over closure.
- *Orthodontic problems*—malocclusion leads to TMJ dysfunction.
- *Emotional problems*—pattern of over behaviors relating to care of teeth, food habits may lead to TMD (temporomandibular dysfunction). Oral habits can cause structural damage or persistent pain. Dysfunction of autonomic nervous system resulting from anxiety and eventually producing structural changes in the end organs.
- *Psychophysiologic theory*—the masticatory muscle spasm is responsible for the signs and symptoms of pain dysfunction syndrome. Persistent myospasm also can cause two other organic changes, namely *degenerative arthritis* which can arise from continued jaw function with condyle in an abnormal position and *contracture* degenerative changes in muscle, that accompanies long term spasm. Spasm can be initiated by muscular over-extension which is caused by:
 - *Dental restoration*—by dental restorations or fixed and removable prosthesis that encroach on the intermaxillary space.
 - *Tooth loss*—by muscular over-contraction which is caused by bilateral loss of posterior teeth.
 - *Denture wearer*—by settling of partial denture or in patients wearing complete denture.
 - *Oral habits*—by muscle fatigue which is caused by oral habits such as clenching or grinding of the teeth.
 - *Dental irritation*—it can be initiated by dental irritation like maloccluding restorations or an over hanging margin.
- *Joint problems*—hypermobility caused by lax ligament may give rise to symptoms of TMJ dysfunction. It has been also shown that patient with steep angulations of articular eminence are more predisposed to the TMJ dysfunction syndrome. Degenerative changes secondary

to chronic para-functional habits or over-closure due to bilateral loss of molar teeth result in TMJ dysfunction syndrome.

Pathogenesis

- During muscle contraction, the energy is released → there is formation and accumulation of lactic acid → which in turn causes changes in osmolality which results in decrease in the pH → it makes the muscle receptor prone to impulse excitation as their critical firing level is impaired → decrease in pH and lactic acid itself causes infusion and effusion of histamine, bradykinin and serotonin and other amines into the area → it causes pathological muscular derangement → these particular areas of muscular derangement are called as 'trigger zones' → it is a hypersensitive area from which impulse bombards the CNS and gives rise to referred pain.

Classification

- *Spasm of lateral pterygoid muscle*—it occurs when the teeth are brought into maximum intercuspation and with extended translatory movement. Pain reduces on biting against a separator (as it prevents the intercuspation required to stretch the spastic muscle). It is accompanied by acute malocclusion expressed as anterior displacement of the mandible.
- *Spasm of elevator muscles*—it occurs when biting and chewing efforts are made and while opening the mouth. Pain is not decreased on biting on a separator and is accompanied by trismus with little or no restriction of excursive movements.
- *Spasm of lateral pterygoid and elevator muscles*—in it, pain occurs while biting, chewing, opening, maximum intercuspation and extended translatory movement. Pain is not affected by biting against a separator, except for some decrease of pain with maximum intercuspation and is accompanied by acute malocclusion, trismus but with little or no restriction of excursive movements.

Clinical Features

- *Age and sex distribution*—it is seen in middle age group with more predilections for women.
- *Onset*—it occurs in episodes of several times a day, at times, with extended symptom free intervals. Usually episodes are seen during increased emotional tension, resulting in increased intra-articular pressure in the joint.
- *Symptoms*—there is masticatory pain which occurs due to myalgia, arthralgia or from both. Pain is localized to preauricular area but can be radiated to temporal, frontal, and occipital region. There is difficulty in chewing and restriction of mandibular excursion. Patient also complains of noise on rubbing, grinding, clicking, and

popping snapping sounds on mandibular movement.

- *Tinnitus*—patient may complain of tinnitus (ringing in ear) or otalgia (pain in ear) or toothache. The reason behind tinnitus is that bilateral molar loss causes distal condylar movement and direct pressure on the eustachian tube with impingement on the auriculotemporal nerve, which is responsible for the ear symptoms, i.e. tinnitus.
- *Hearing loss*—it may cause irritation of the chorda tympani nerve, resulting in partial or total hearing loss.
- *Signs*—restriction of opening and protrusion may be accompanied by deflection of the mandibular incisal pathway. There is also soreness of muscle, when palpated. Myofascial trigger zones are stimulated by pressure and produce referred pain.
- *Myospasm*—muscle splinting can lead to CNS induced muscular contraction. In some cases, myositis may also occur.
- *Other features*—oral or para-functional habits, such as bruxism, present as indentation on lateral borders of the tongue, ridging of the buccal mucosa and extensive attrition of teeth.
- *Involvement of various masticatory muscles and their clinical effects.*

Muscle	Pain refers to	Clinical effect
Temporalis	Temple, maxillary teeth TMJ	Restriction of mandibular opening, ipsilateral deviation of mandible, deviation of interocclusal space
Masseter	Mandible, maxillary molar, TMJ, ear	Same as above
External pterygoid	TMJ	Contralateral deviation of the mandible, protrusion of condyle, acute malocclusion
Internal pterygoid	TMJ, retromandibular area, tongue	Restriction of mandibular movements, contralateral deviation of mandible

Laskin's Diagnostic Criteria for MDPS

- *Four cardinal signs*
 - *Unilateral pain*—it is generally a dull ache in nature felt in the ear or the preauricular area or at the angle of the mandible. The pain is more often worse on arising in the morning or relatively mild in the morning, but gradually becomes worse as the day progresses.
 - *Muscle tenderness*—the most frequent areas are the neck of mandible and in the region distal and superior to the maxillary tuberosity.
 - *Clicking*—clicking or popping noise in the TMJ.
 - *Limitation of jaw function*—limitation of jaw function or deviation of the mandible on opening.

- *Negative characteristics*
 - *No radiographic evidence*—absence of radiographic or biochemical evidence in the TMJ.
 - *No tenderness*—lack of tenderness in TMJ area, on palpation via the external auditory meatus.

Management

Education regarding present illness

- *Re-educate* the patient about the mechanism involved and factors causing or aggravating the disorder. The patient is convinced about the disease status.
- *Reassurance*—the patient is assured of the benign character of the myospasm. The patient should be explained that there will not be any residual deformities after the treatment.
- *Suggestion*—may act as a powerful tool in the treatment. The calm attitude of the clinician and reassurance given suggests the patient that recovery is bound to occur. Let the suggestions act in a natural quiet fashion rather than trying to use it in some spectacular ways.

Muscle relaxation techniques and muscle exercise

- *Tongue exercise*—the patient is asked to keep his tongue as posteriorly as possible, in contact with the palate, while keeping the mandible in a retruded position. In this exercise, mainly rotatory movements are performed.
- *Mouth opening exercise*—the patient is asked to open his mouth slowly, rhythmically, within the pain limit, 10 times in succession. Next, the patient is asked to repeat such exercises with the addition of one modification, i.e. voluntary resistance.
- *Voluntary resistance*—this technique is based on the physiologic principle of reciprocal muscle inhibition (reflex inhibition), e.g. on depressing the mandible, the contraction of the depressor muscle group causes an inhibition of the elevator muscles. Thus, if a muscle is subjected to a greater degree of inhibition, its relative relaxation will increase proportionately. This muscular relaxation will increase circulation to the antagonist system. It is during this period of active reflex inhibition, that the noxious stimuli present in and about the trigger zone can be eliminated. Resultantly, the inter-muscular blood vessels get dilated. As more muscle fiber areas relax and fewer remain in spasm, the patient evidences gradual, painless and rhythmic mandibular movements.

Pharmacotherapy

- *Pain control*—it is usually achieved by analgesics. Most widely used analgesic is salicylates, the commonest being aspirin.
- *Tranquilizers*—there are major and minor tranquilizers. The tranquilizers used in major psychoses, like

phenothiazine derivatives, having calming effect in anxiety states associated with neurotic personality. It relieves tension, fear and produces a sense of well being.

- *Antidepressants*—they are mood elevators like monoamine oxidase inhibitor, lithium carbonate and caffeine.
- *Sedative*—sedatives are the drugs that reduced excitement.
- *Hypnotic*—hypnotics produce sleep which resembles natural sleep.

Biofeedback

- *EMG biofeedback device*—the most commonly used biofeedback device is electromyographic (EMG) biofeedback machine. The device senses cutaneous electrical output. Every time the muscle contracts and electrical current is given off, which diffuses outward and may reach the skin. Current from the muscle nearer to the skin is much earlier to detect than current from deeper muscles. Muscle gives off a specific frequency of electrical output. The biofeedback elaborately gauges the amount of amperage and voltage. This method is non-invasive and painless.
- *Temperature biofeedback*—the temperature biofeedback accounts of placing a delicate thermometer on the skin. The temperature is measured in tenth of degree centigrade. The higher the temperature, the higher the pitch of sound. When there is poor circulation, individual can increase the temperature by this method.

Bruxism prosthesis

- *Soft mouth guard*—it is a bruxism guard made up of soft resin. It may be fashioned for either upper or lower teeth and is designed even for centric contact of all the teeth.
- *Anterior occlusion prosthesis*
 - *Lucia jig*—it consists of a smooth acrylic rim placed on the lingual aspect of the upper incisors. It should be complete enough to provide complete posterior disocclusion.
 - *Anterior bite plate*—this appliance does not produce posterior contact and hence can be used in patients with bruxism. It should not be used for more than one or two weeks as than it may raise the bite.
- *Mandibular posterior coverage*—it covers the posterior teeth of the mandible, when the jaws are closed. It causes bilateral, single point contact on the most distally placed tooth. It is made up of acrylic or metal.
- *Complete maxillary or mandibular coverage*—it covers all occlusal surfaces in either maxillary or mandibular arch. By gripping all the teeth in one arch, it acts as a true splint, preventing the tooth movement and minimizing the number of posterior teeth occluding. In addition, anterior guidance will provide posterior disocclusion in all eccentric movements.

Psychotherapy

- *Stress impasse stages*—the stress impasse stages in the TMJ syndrome are characterized by three distinct steps based on the evolution and progression of the disease.
 - *Stress impasse I*—conflict from personality fixation, usually in adolescents, chronic symptoms develop (bruxism, clenching, grinding).
 - *Stress impasse II*—onset of acute symptoms (pain, jaw clicking).
 - *Stress impasse III*—severe symptoms resulting in TMJ damage.
- *Treatment plan*—the treatment plan includes a three stage programme, which will include basic relaxation exercises, sleep preparation and containment strategies for worry, grief and guilt.

Physical medication

- *Hot packs*—the one of moist heat is advantageous. Heat should be applied directly to the trigger zones. Temperature changes can alter the amount of vasodilation substance held within the tissue spaces. The patient should be instructed to place hot fomentation over a particular area for not less than 10 minutes, preceding the therapeutic exercises. Woolen cloth soaked in boiling water is usually used.
- *Massage*—it provides relief from muscle spasm and also improves the local circulation, by reducing the local biogenic amines.
- *Diathermy and ultrasound*—this is a more effective method for the treatment of myospasm of deeper muscles. It produces heat and thus improves the local circulation.
- *Electrical stimulation (TENS)*—tetanizing and sinusoidal current is used in this therapy. It is another effective way to break acute painful muscle spasms. Electrodes are placed over the most painful areas and then tetanizing current is turned on gradually, until good contraction of the affected muscle has been obtained. Current should not exceed the tolerance level. The current is left for 120 minutes. Then with the electrode remaining in the same position, the tetanizing current is switched off and sinusoidal current is gradually applied, till good contraction is obtained.
- *Oral myofunctional therapy* is a program to re-educate the muscles of the mouth and face to work in a balanced relationship. It includes breaking of a habit (Lip biting, tongue thrusting), proper feeding of infant and proper balancing of environmental forces. The appliances used are activator, bionator, Fränkel's appliance and oral screen.

Applied kinesiology

- *What is it*—dental Kinesiology is the study of motion and function of the jaws, oral musculature, the accompanying neurological, vascular and other support system network and the impact these muscle functions

and neurological dynamics have on dental and systemic health. Its basic application is for reduction and prevention of muscle spasm.

- Muscles are tested to identify the weak muscle. The clinician then uses various kinesiologic procedures to increase the stress resistance of the tensed antagonist muscle to a more optimum level.

Anesthesia

- *Muscle and fascia (trigger points)*—injection of local anesthetic agent into the painful trigger zones is therapeutically efficient. It permanently terminates the pain-spasm-pain cycle. Local anesthesia breaks the cycle by blocking the nerve impulse conduction and by eliminating pain from that area. Local anesthesia should be used without vasoconstrictor component, i.e. adrenaline. It is important to first isolate the trigger zone. Then, it is gently probed to reach for the most hyper-sensitive tissue. Once defined, slow infiltration of anesthetic solution is deposited. The patient is then instructed to begin a series of exercises.
- *TMJ (intracapsular and extracapsular)*—many a times, xylocaine is used in conjunction with injection of hydrocortisone for the intra-articular injection into the TMJ. 0.5 ml of 0.5% xylocaine is generally used.
- *Refrigerated spray*—vapocoolant spray, such as ethyl chloride or fluoromethane, is used to reduce muscle spasm by causing counter irritation. Since pain and temperature sensation travel by the same spinal pathways, it is theorized that response to pain impulse originating from the trigger zone, is completely inhibited and thus, inhibition of the vicious cycle is achieved. The spray is applied with a slow sweeping motion over the trigger points. Skin frosting should be avoided.

Other therapies

- *Hypnotherapy*—here, the patient's cooperation is essential and he should follow the suggestions of the hypnotist. It is an individual's belief that aids him in following the suggestions of the hypnotist. By this method, muscle relaxation is achieved.
- *Acupuncture*—it is a simple, effective and conservative pain control modality. It is accomplished by inserting needles into appropriate joints, at appropriate surfaces called as meridians. These meridian points are generally associated with major nerves, arterial pathways, joints and fascial planes. Rotating a sharp, small needle between the fingers rapidly and pressing it through the skin rapidly, in the acupuncture point provides relief from pain. The patient may feel soreness, distension, heaviness or numbness, if it is placed into so called acupuncture point. However, it should be kept in mind that this therapy is used only to give relief from pain and it will not remove the basic cause.

- *Surgery*—various surgical procedures like eminectomy, zygomectomy, menisectomy, high condylectomy with material interposed between the articulating surfaces and orthoplasty plus menisectomy are most frequently advocated.
- *Restoration*—faulty overfilled restorations should be removed to avoid interceptive contact. Sometimes, complete mouth rehabilitation is required to achieve centric occlusion.
- *Prosthodontic therapy*—it has been shown that overclosure due to bilateral loss of posterior teeth can cause TMJ pain. In such cases, bilateral fixed or removable replacement should be done to achieve normal vertical dimension.
- *Orthodontic therapy*—it is indicated in cross bite and traumatic bite. In patient with parafunctional habits, such as bruxism, night guard appliance should be given.
- *Orthognathic therapy*—when there is skeletal discrepancies causing malocclusion, orthognathic surgery is indicated.

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26

Salivary Gland Disorders

Development of Salivary Gland

The three major sets of salivary gland—the parotid, submandibular and sublingual originate in a uniform manner by oral ectodermal epithelium buds invading the underlying mesenchyme.

- *Parotid gland*—the parotid gland develops in the lateral aspect of stomodeum. Parotid gland buds are the first to appear at the 6th week of intrauterine life, on the inner aspect of cheek, near the angles of mouth and grow back towards the ear. In the parotid and ear region, the epithelial cord of cells branches and canalizes to provide the acini and ducts of the gland. The duct and acinar system are embedded in a mesenchymal stroma, get organized into lobules and become encapsulated. The parotid gland duct, although repositioned, traces the path of embryonic epithelial cord in the adult.
- *Submandibular gland*—it develops in the floor of stomodeum. Submandibular gland buds appear in the 6th week of intrauterine life as a grouped series, forming epithelial ridges on the either sides of the midline, in the floor of the mouth. An epithelial cord proliferates back into the mesenchyme beneath the developing mandible to branch and canalize, forming the acini and ducts of submandibular gland. The mesenchymal stroma is separated off the parenchymal lobules and provides a capsule for the gland.
- *Sublingual gland*—it also develops in floor of stomodeum. It arises in the 8th week of intrauterine life, as a series of about ten epithelial buds, just lateral to the submandibular gland analogue. These branches and canalize to provide a number of ducts opening independently beneath the tongue.
- *Minor salivary gland*—minor salivary gland begins their development during the third month of life. These glands arise from the oral ectodermal and endodermal epithelium. *Labial glands* arise during the 9th week of intrauterine life and are morphologically mature by 25th week.

Classification of Salivary Glands

- *Exocrine glands*—these are the glands whose products are carried away by the ducts leading from the gland.
- *Mesocrine glands*—the secretory products pass through the cell walls losing the cytoplasm.
- *Serous salivary gland*—the salivary glands which produce a thin watery secretion are called as serous.
- *Mucus salivary gland*—these produce a thick, viscous substance called mucus.
- *Seromucus salivary gland*—salivary glands which produce serous and mucus in varying quantities.
- *Major salivary gland*—these are large salivary glands which are located outside the oral cavity and convey their secretions through their ducts.
- *Minor salivary gland*—these are smaller salivary glands confined to the mucus and submucus coat of the oral cavity.

Major Salivary Glands

Major salivary glands are parotid, submandibular and sublingual.

Parotid Gland

- *Location*—it comes from the word para—around and otic—ear. It is like an inverted flattened pyramid. It is the largest of the salivary glands weighing about 15 grams each. It lies between the mastoid process and vertical ramus of the mandible. The bulk of the parotid gland is situated in the retromandibular fossa (**Fig. 26-1**).
- *Shape*—it is wedge shaped, with the broad edge of the wedge lying subcutaneously and the apex lying deep between the parotid fascias. It is divided into superficial and deep lobes by the facial nerve and its branches. It forms an irregular lobulated yellowish mass, lying below the external acoustic meatus, between the mandible and

the sterno-cleidomastoid. A small part of it, more or less detached lies between the zygomatic arch superiorly and the parotid duct inferiorly is named '*accessory part*' of the gland.

- *Stensen's duct*—the parotid duct which is called as '*Stensen's*' duct is about 5 cm long and has thick walls. It emerges from the substance of the gland to course anteriorly until it reaches the anterior border of the masseter muscle at the point of upper and middle thirds. When it crosses the masseter muscle it receives the duct of the accessory lobe. Around the border of the masseter muscle, the duct turns sharply medially, often embedded in a furrow of the protruding buccal fat pad. In its medial course, the duct reaches the outer surface of the buccinator muscle, where it perforates in an oblique direction anteriorly and medially. It then runs for a short distance obliquely forward, between the buccinator and mucus membrane of the oral cavity and opens on the oral surface of the cheek, opposite the upper second molar.
- *Blood supply*—parotid gland is supplied by the external carotid artery and its branches near the gland.
- *Lymphatic drainage*—drains first to the parotid nodes and from there to the upper deep cervical nodes.
- *Nerve supply*—it is supplied by auriculo-temporal nerve, plexus around the external carotid artery and greater auricular nerve.

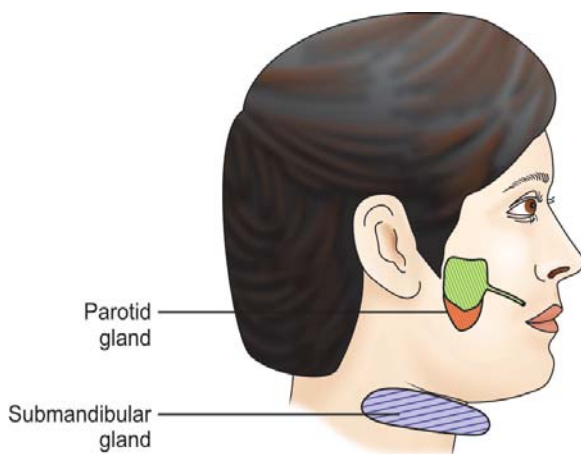


Fig. 26-1: Location of parotid and submandibular gland.

Submandibular Gland

- *Shape and size*—it is a round biconvex salivary gland situated in the anterior part of the digastric triangle. It is irregular in form and about the size of a walnut.
- *Location*—it is enclosed by two layers of deep cervical fascia. The inner surface of the submandibular gland is in contact with stylohyoid, digastric and styloglossus muscle, posteriorly and with the hyoglossus and posterior border of the mylohyoid muscle, anteriorly.

- *Wharton's duct*—the submandibular duct which is called as '*Wharton's duct*', is about 5 cm long and its wall is much thinner than that of parotid duct. It emerges from the middle of the deep surface, of the superficial part, of the gland. It runs forward, beneath the deep part of the gland, between the mylohyoid and hyoglossus muscle. It runs further forward between the medial surface of the sublingual gland and the genioglossus muscle and finally ends by opening into the summit of the sublingual papilla, situated in the floor of the mouth, on the side of the frenulum. The last few millimeters of the duct are often slightly widened.
- *Arterial supply*—the arteries supplying the submandibular gland are derived from the lingual and facial branches of external carotid artery.
- *Venous drainage*—it drains into facial and lingual vein.
- *Nerve supply*—its nerve supply is from the branches of submandibular ganglion through which it receives fibers from chorda tympani.
- *Lymphatic drainage*—passes to the submandibular lymph node.

Sublingual Gland

- *Location*—it lies above the mylohyoid and below the mucosa of the floor of mouth. It is medial to the sublingual fossa of the mandible, on either side of the symphysis menti and lateral to genioglossus muscle. It has about 15 ducts which open directly into the floor of mouth.
- *Bartholin's duct*—the duct of sublingual gland is called as '*Bartholin's duct*'. They are eight to twenty in number. Some of the smaller sublingual ducts open into the sublingual fold, in the floor of the mouth, on either side of lingual frenum. Some open into the submandibular duct and others unite to form the "*principle sublingual duct*" which opens in the floor.
- *Blood supply*—it is supplied by sublingual and submental arteries.
- *Nerve supply*—it is supplied by lingual and chorda tympani nerve.
- *Lymphatic drainage*—it passes to the submandibular lymph nodes.

Minor Salivary Glands

The minor salivary glands are located beneath the epithelium in almost all parts of the oral cavity. These glands usually consist of several small groups of secretory units, opening via short ducts directly into the mouth. The function of minor salivary glands is not really to produce saliva for mixing with food, but to secrete minor amounts of saliva onto the mucosal surface to keep the mucosa moist.

- **Labial glands**—they are found on the submucosa of upper and lower lips and opens onto the inner surface (Fig. 26-2). They are more numerous in the areas close to midline. The labial glands classically have been described as mixed, consisting of mucus tubules with serous demilunes.

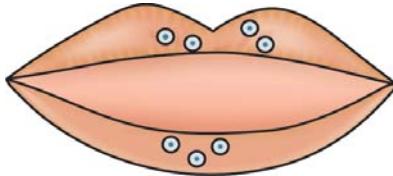


Fig. 26-2: Location of labial gland.

- **Buccal glands**—they are found in inner cheek region. In the anterior part of the cheek, they are sparse and rather widely and irregularly spaced. They are usually described as continuation of labial glands, as they have similar structure. A group of these glands is situated at the lower posterior corner of the cheek which is called as the 'molar or retromolar glands'.
- **Palatine glands**—the palatine glands are located in soft palate and posterolateral part of hard palate (Fig. 26-3). They are pure mucus in nature. The excretory ducts may have an irregular contour and opening of the ducts in the palatal mucosa is large.

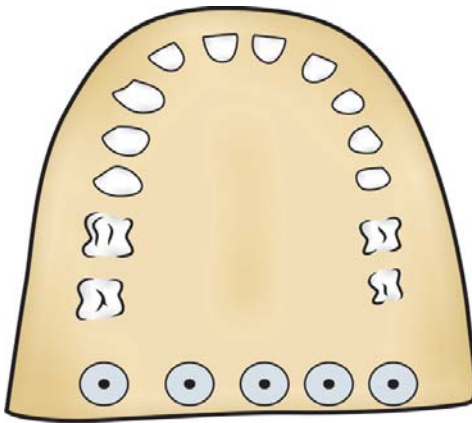


Fig. 26-3: Location of palatine gland.

- **Glossopalatine glands**—these are pure mucus glands. They are principally located in the region of the isthmus of the glossopalatine fold but may extend from the posterior extension of the sublingual gland to the glands of soft palate.
- **Gland of Blandin's and Nuhn's**—these are the anterior lingual glands. They are located near the apex of the tongue and are chiefly mucus in nature. They are covered by thin mucus membrane of the inferior surface of the tongue. It is compact package of smaller glands that open with several ducts on the inferior surface of the tongue.

- **Gland of von Ebner**—lingual glands of von Ebner are posterior lingual serous glands, located between the muscle fibers of tongue, below the vallate papillae.
- **Posterior lingual glands**—posterior lingual glands are located lateral and posterior to the vallate papillae and in association with the lingual tonsils. They are purely mucus in character.
- **Incisive gland**—the incisive glands are a small group of glands found on the floor of the oral cavity close to the insertion of the lingual frenum, behind the lower incisors (Fig. 26-4).

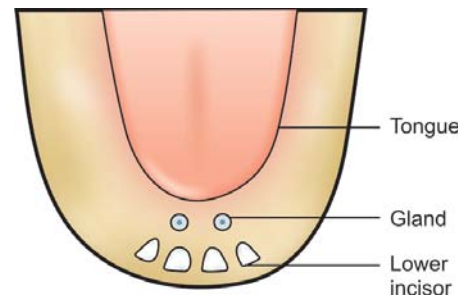


Fig. 26-4: Location of incisive gland on floor of mouth.

Saliva

Composition of Saliva

- **Source of saliva**—saliva is secreted by salivary glands: viz the parotid, submandibular and sublingual glands. These glands produce approximately 1.5 liters of saliva daily: 70% is from the submandibular glands, 25% is from the parotid glands and 5% is from the sublingual glands. Minor salivary glands located on the palate, buccal mucosa and tongue also produce modest amounts of saliva.
- **Normal stimulated flow of saliva**—the normal stimulated flow for different ages can be calculated with following equation:

$$0.78 \times \text{age} + 5.6 = \text{stimulated flow/15 minutes.}$$

5.6 – it is estimated stimulated flow of the infants

- **Composition of saliva**—saliva is slightly cloudy in appearance due to the presence of cells and mucin. Saliva is slightly acidic and the pH varies from 6.7 to 7.4, specific gravity is 1.002 to 1.012. The resting flow rate of saliva, on an average is about 0.3 to 0.4 ml/min. Saliva contains about 99.5% of water and 0.5% of solid.
- **Solid**
 - **Cellular constituents**—consist of yeast cells, bacteria, protozoa, polymorpho-nuclear leukocytes and desquamated epithelial cells.

- *Inorganic ions*—major (Na^+ , K^+ , Cl^- , HCO_3^-) and minor (Ca^{++} , Mg^{++} , HPO_4^- , I , Scn^- and F^-).
- *Secretory proteins and glycoproteins*—various enzymes, large carbohydrate rich protein or mucin, antibacterial substance, group of proteins involved in enamel pellicle formation and calcium phosphate.
- *Serum constituents*—albumin, blood clotting factor, B_2 microglobulin and immunoglobulin.

Collection Technique

Parotid Collector

- *Composition*—it was developed by *Lashley* in 1916. It makes possible the collection of parotid fluid uncontaminated by the oral content. It is composed of two concentric circles and is made of either plastic or metal.
- *Technique*—the center circle is designed to fit over the opening of Stensen's duct and is connected to a graduated collecting tube. The outer concentric circle is attached to a rubber bulb, which exhausts air from the outer circle, when collector is held in place and draws the cheek surrounding the opening of Stensen's duct into it.
- *Disadvantage*—due to different anatomy, it cannot be used in submandibular and sublingual gland.

Segregator

- *Composition*—it is developed by *Schneyer* which allows the collection of submandibular and sublingual gland. It is made up of plastic or metal. It must be constructed for each individual on the stone model. In it, preformed basic plastic collector is utilized.
- *Technique*—this plastic is covered with rubber base impression material and placed on the floor of mouth beneath the tongue. In 5 minutes, impression can be removed. A recess is then made in the impression, over the opening of Wharton's duct and plastic collecting tube is attached. The collector stays in position when the patient places his tongue against the lingual surface of the lower incisors.
- *Advantage*—it may be sorted and reused for the individual patient. It collects saliva from sublingual and submandibular ducts.

Collection of Whole Saliva

- *Stimulation of saliva*—for collection of whole saliva, stimulation of saliva is to be done by asking the patient to suck on sour candy or sour grapes. You can also ask patient to chew on cimer paraffin or rubber bands. A more standardized procedure is to swab a solution of 2% citric acid on the back and side of tongue at 15 seconds

intervals. Secretion can be obtained by draining, spitting and suction.

- *Draining*—in this, the subject is placed with his head inclined forwards, so that saliva will collect in the anterior floor of the mouth. The position of the patient allows the saliva to flow over the lip, where it is collected by funnel.
- *Spitting*—in this, the subject actively spits into the collecting funnel at regular intervals.
- *Suction*—saliva ejector is applied orally in the area of lower incisors and the aspirated fluid is collected after the patient has remained quits for fixed time periods.

Function of Saliva

- *Digestive function*—it participates in digestion by providing fluid environment for solubilization of food and taste substance, through action of enzyme amylase and lipase. It helps in formation of food bolus, which is readily chewed.
- *Lubricating*—it keeps the oral tissues moist and facilitates swallowing and speaking. Mucus glycoproteins provide lubricant for the movement of oral tissues against each other and protection from chemical and thermal insults.
- *Protection of teeth*—it helps to protect the teeth from dental caries by means of, both, cleansing and buffering action of saliva. It also controls the calcium and phosphate concentration in the saliva and around the teeth.
- *Protection from injury*—it dilutes hot or irritant substance and thus prevents injury to mucus membrane.
- *Maintenance of mucus membrane integrity*—the salivary mucin possesses rheological properties which include low solubility, high viscosity, elasticity and adhesiveness which enable them to concentrate on the oral mucosal surface. They provide an effective barrier against desiccation and environmental insult. Salivary mucin binds water effectively and hence serves as natural 'water proofing' and helps to maintain these tissues in a hydrated state. The glycosylated mucins of saliva are resistant to proteolysis. This maintains the integrity of mucus membrane by neutralizing the bacterial and PMN proteases.
- *Antibacterial properties*—several salivary substances are capable of inhibiting the growth of microorganisms. They are glycoproteins, antibacterial protein, i.e. lysozyme, immunoglobulin IgA, IgG, IgM and lactoferrin.
- *Debridment and lavage*—the physical flow of saliva, augmented by muscular activity of lips and tongue, effectively removes a large number of potentially harmful bacteria from the teeth and mucosal surface.
- *Anti-carcinogenic activity*—mucins limit the penetration of variety of potential irritants and toxins in foods, beverages as well as potentially hazardous agents from tobacco smoke and other sources.

- *Soft tissue repair*—presence of nerve growth factor and epidermal growth factor in submandibular saliva accelerates wound healing.
- *Maintenance of tooth integrity*—after the eruption the crown, the tooth is fully formed morphologically, but is crystallo-graphically incomplete. Interaction via diffusion of ions such as calcium, phosphorus, magnesium, fluoride as well as other trace components into the surface of enamel, increases the surface hardness, decreases permeability and increases the resistance to caries. Acquired pellicle, which covers the tooth surface after it begins to function in oral cavity, is made up of salivary proteins and lipids.
- *Maintenance of pH*—it helps in maintenance of neutral pH in oral cavity and in esophagus. The salivary bicarbonates are responsible for maintenance of pH. Saliva also neutralizes the acids produced in bacterial plaque because of bacterial metabolism of carbohydrates.
- *Excretory functions*—many drugs as well as alcohol are excreted into saliva which could theoretically serve as a route of elimination.
- *Water balance*—salivary glands are a part of the control system for maintaining an appropriate level of hydration. Thirst and the need for fluid intake are usually originated by a dry mouth, which activates receptors in the oral cavity.
- *Hormonal function*—polypeptide hormones known as epidermal growth factor, is identical with human urogastrone which is found in high concentration in urine. It is also found in submandibular and parotid saliva.
- *Bimanual palpation*—palpation should be performed in the lateral surface of the mandible and on the soft tissues inferior and medial to the angle. Bimanual palpation with the patient's mouth closed: with the masseter muscle relaxed, this method can be readily performed from the side or behind. Insertion of the index finger along the teeth to the most posterior location in the cheek with application of lateral pressure against the examining thumb on the face, this palpation can be performed.
- *Examination of Stensen's duct*—Stensen's duct orifice should be identified adjacent to the upper molar teeth as a small fold or soft tissue flap in the buccal surface. The mucosa in the area should be dried with cotton sponge and the duct orifice observed for secretion while the gland is milked by applying firm pressure. The pressure is applied first in the posterior aspect, beneath the auricle and moving the palpating finger anteriorly and inferiorly, along the course of Stensen's duct. Clear, colorless secretion that is sufficiently fluid to flow rapidly should be observed emanating from the duct orifice.

Submandibular Glands

- *Location of swelling*—submandibular salivary gland enlargement is characterized by medial and inferior extension, the later resulting in discontinuity if the tissue contours at the inferior border of mandible.
- *External palpation*—external palpation should start with the finger extending towards the midline and the thumb on the body of the mandible. Pressure is exerted, both, superiorly and laterally and the finger is gradually moved beneath the inferior border of the mandible.
- *Palpation*—the patient should be advised to relax the tongue during palpation. Enlarged lymph nodes, except those involved with malignancy, will invariably move laterally with the examining finger, while the submandibular salivary gland will remain stationary.
- *Bimanual palpation*—after this, bimanual palpation should be performed; the examiner placing the 2nd finger of one hand into the floor of the mouth beneath the tongue, the patient resting his teeth on the examiners fingers, while the fingers of the other hand are placed as previously described, that is on the skin beneath and medial to the body of the mandible. While pressure is exerted inferiorly intraorally by the finger, the other hand moves superiorly and laterally so that all organs come between the examiner's two hands.

Sublingual Salivary Glands

- *Location*—these are located in the floor of the mouth in the region of the middle third of the tongue, closely applied to its medial musculature attachment. The ductile pattern is extremely variable, frequently communicating with the mouth through a series of small orifice, rather than by means of small collecting ducts.

Examination of Salivary Glands

Minor Salivary Glands

- *Technique*—the minor salivary glands can be examined by simply drying the lower lip mucosa with cotton sponge, after eversion using thumb and index finger. Careful observation for the appearance of small globules of fluid emanating from minor gland duct orifice should be done. In situations where atrophy or significant reduction in salivary secretion has occurred, such as in Sjögren's syndrome, the duct orifices will be marked by small red spot and secretion will not be observed.

Major Salivary Glands

Parotid Glands

- *Location of swelling*—swelling of the parotid gland is usually associated with lateral extension of the lobe of the ear and difficulty in opening the jaw widely.

Diagnostic Tests of the Salivary Glands

- *Sialography*—it is defined as a method of studying the salivary gland and the alveoli of the parotid and submandibular salivary gland radiographically. It is done by injecting radiopaque contrast medium or a retrograde injection of a radiopaque material into the duct system of a salivary gland and the study of its distribution by roentgenogram.
- *Salivary gland scanning (scintigraphy)*—it is used for studying the glandular parenchyma. The salivary gland tissues take up compounds of periodic group VII elements such as iodine, bromine and technetium. It provides indication of salivary gland functions and allows bilateral comparisons and images of all four major salivary glands at the same time.
- *Ultrasonography*—it involves the transmission of energy into the salivary tissues, receiving of the energy after it has been reflected by the tissues and recording it so that it can be presented for interpretation. It is also useful in identification of radiolucent stone. Different echo signals are obtained from different tumors.
- *Computerized tomography*—it demonstrates small differences in soft tissues X-ray examination and distinction between gland and the adjacent soft tissues is greatly improved. It is usually done in cases of discrete swelling, both intrinsic and extrinsic to the salivary gland.
- *Arteriography*—it will define the vasculature of the tumor but also delineates the origin of vascular supply.
- *Biopsy*—it has been the most significant advancement in diagnosis and appropriate treatment of major salivary gland tumors.
- *Flow rate studies*—comparative study of flow rate from major salivary glands is done over a time period. It provides information about salivary gland functions.
- *Magnetic resonance imaging*—it is useful in discrete swelling of salivary glands and provides excellent soft tissue details. It readily enables differentiation between the normal and abnormal.

Classification

Developmental disorders

- Aberrancy
- Aplasia and hypoplasia
- Hyperplasia
- Atresia
- Accessory ducts
- Diverticuli
- Congenital fistula

Functional disorders

- Sialorrhea
- Xerostomia

Obstructive disorders

- Sialolithiasis
- Mucus plug
- Stricture and stenosis
 - Foreign bodies
- Extra-ductal causes

Cyst

- Mucocele
- Ranula

Asymptomatic enlargement

- Sialosis
- Allergic
- Associated with malnutrition and alcoholism

Infection

- Viral infection
- Bacterial infection
- Mycotic infection

Autoimmune disorders

- Sjögren's syndrome
- Mikulicz's disease
- Uveoparotid fever
- Recurrent non-specific parotitis

Developmental Disorders of Salivary Gland

Aberrancy

It is defined as the situation in which the salivary gland tissue develops at a site where it is not normally found. It is also called as *ectopic salivary gland*.

Ectopic salivary tissue can develop anywhere within the territory of the first and second branchial arches, in the lateral neck, pharynx or middle ear.

Clinical Features

- *Site*—true aberrant salivary glands are most frequently reported in the cervical region, near the parotid gland or body of the mandible. The salivary gland tissue in the mandible is found posterior to the first molar and often has a small communication with a major salivary gland.
- *Developmental lingual salivary gland depression*—the aberrancy of the salivary gland tissue represents only an extreme example of the condition known as the 'developmental lingual mandibular salivary gland depression'. It is the developmental inclusion of the

glandular tissue within or more commonly, adjacent to the lingual surface of the body of the mandible, in a deep well circumscribed depression. It was first described by Stafne in 1942 and hence referred to as 'Stafne's cyst'.

- *Salivary tissue in neck*—salivary tissue is regularly found in lymph nodes within the neck and can be mistaken for metastatic disease, if found in a neck dissection specimen.
- *Gingival salivary gland choristoma*—ectopic salivary gland tissue has been reported to occur in the gingiva, where it may be described as 'gingival salivary gland choristoma'.
- *Clinical significance*—they may become site for development of a retention cyst or neoplasm.

Aplasia and Hypoplasia

Aplasia or agenesis is the congenital absence of the salivary gland. It was first described by Gruber in 1885. Aplasia may occur in association with other developmental abnormalities such as atresia of lacrimal puncta and congenital malformation of temporomandibular component.

Causes

- *Ectodermal origin*—Macdonald suggested ectodermal origin for this anomaly.

Clinical Features

- *Sites*—any one of the glands or group of glands is missing, either unilaterally or bilaterally. Hypoplasia of salivary gland is rare but hypoplasia of parotid gland has been reported to be present with Melkersson-Rosenthal syndrome.
- *Symptoms*—patient complains of xerostomia, which may be so severe as to necessitate the constant sipping of water throughout the day and particularly, during meal times.
- *Significance*—the lack of saliva results in rampant dental caries and early loss of deciduous and permanent teeth.
- *Appearance of oral mucosa*—the oral mucosa appears dry, smooth, or sometimes pebbly and shows a tendency for accumulation of debris.
- *Signs*—patients exhibit characteristic cracking of lips and fissuring of the corners of mouth.

Radiological Features

- *CT features*—it is sometimes observed that unilateral gland is missing (Fig. 26-5) or is very small compared to the contralateral side with congenital or posterior reasons. As they are mostly asymptomatic, the condition is mostly diagnosed by chance.



Fig. 26-5: Hypoplasia of the unilateral parotid gland (Plain CT). Size of the left parotid gland (yellow arrow) is remarkably smaller compared with normal size of the right parotid gland (red arrow) (Courtesy M Shimizu).

Diagnosis

- *Clinical diagnosis*—decreased production of saliva without any other visible clinical anomalies this disease should be suspected.
- *Radiological diagnosis*—Computed tomography will be able to diagnose this lesion.

Management

- *Maintenance of oral hygiene*—institution of scrupulous oral hygiene in an attempt to decrease dental caries and preserve the teeth as long as possible.

Hyperplasia of Salivary Gland

Hyperplasia is the increase in the size of salivary gland.

Causes

- *Hormonal disorders*—endocrine disorders and menopause.
- *Metabolic disorders*—gout, diabetes mellitus.
- *Autoimmune*—Sjögren's syndrome, Waldenström macroglobulinemia.
- *Syndrome*—aglossia-adactylia syndrome, Heerfordt's syndrome and Felt's syndrome.
- *Miscellaneous*—hepatic disease, starvation, alcoholism, inflammation, benign lympho-epithelial lesion, adiposity, hyperthermia, oligomenorrhea and certain drugs.

Clinical Features

- *Prevalence*—it is more common in minor salivary glands of the palate.

- *Size and site*—palatal gland hyperplasia appears as small localized swelling of varying size, measuring from several millimeters to 1 cm, usually on the hard palate or at junction of hard and soft palate.
- *Surface*—the lesion has an intact surface and is firm, sessile and normal in color.
- *Symptoms*—it is usually asymptomatic.

Radiological Features

- *CT features*—Computed tomography will diagnose this condition.

Diagnosis

- *Clinical diagnosis*—it is not specific.
- *Radiological features*—computed tomography will diagnose this condition.

Management

- *Excision for microscopic examination*—as it cannot be differentiated from minor salivary gland tumors, it becomes essential to excise it for microscopic examination.

Atresia

It is the congenital occlusion or absence of one or two major salivary gland ducts. Usually the submandibular duct in the floor of the mouth fails to cannulate during embryological development. The newborn infant presents, within 2 or 3 days of life, with submandibular swelling on the affected side due to the presence of a retention cyst. It may produce a relatively severe xerostomia.

Accessory Duct

An accessory parotid lobe is the most common developmental anomaly. It occurs in as many as 20% of subjects. Its position is constant, arising from the horizontal component of the parotid duct as it crosses the masseter muscle. Its importance lies in the fact that any of the diseases that can affect the salivary glands, may involve the accessory lobe and lead to diagnostic confusion, as the possibility is not considered. This is because the symptoms and signs are not within the normal anatomical territory of the parotid. Presence of additional duct in some salivary glands has been reported.

Diverticuli

They are small pouches or out pocketing of the ductal system of one of the major salivary glands. Their presence leads to recurrent episodes of acute parotitis.

Congenital Fistula

Patients with branchial cleft anomalies usually present with unilateral, painless swelling in the region of parotid. Rarely, they are bilateral. They form sinus tracts either in the crease behind the pinna or in front of tragus. They discharge saliva intermittently. Abscess formation, due to secondary infection, may occur. Complete surgical excision of the sinus tract is essential. The dissection is often very extensive and full dissection of facial nerve may be required.

Developmental Salivary Gland Defect

It is also called as '*static bone cavity or cyst*', '*Stafne's cyst or defect*', '*lingual mandibular bone cavity*', '*lingual cortical mandibular defect*', and '*latent bone cyst*'. It is the developmental inclusion of glandular tissue within or more commonly, adjacent to the lingual surface of the body of mandible. It was recognized by Stafne in 1942. Mandible develops around the lobe during development. As it remains stable in size, it is called as '*static bone cyst*'.

Clinical Features

- *Incidence and sex*—it is rare. Males are affected more commonly than females.
- *Site*—it is located in the posterior body of the mandible.
- *Sublingual gland bony defect*—in some cases cortical defect can also occur in anterior region. These defects are related to sublingual gland.
- *Symptoms*—it is asymptomatic and only diagnosed on radiographical examination.
- *Sign*—sometime a notch or depression can be palpable on clinical examination in the posterior area.

Radiographic Features

- *Site*—it is found below the mandibular canal and above the inferior border of mandible, just anterior to the angle of jaw and below and just posterior to third or second molar (**Fig. 26-6**).
- *Size and shape*—round or ovoid radiolucency that will vary in size from 1 to 3 cm in diameter. It is occasionally bilateral.
- *Borders*—it is well defined by dense radiopaque border that is the result of the rays passing tangentially through the relatively thick wall of depression.
- *Sublingual bony defect*—in some instances, round or ovoid radiolucency may occur in the anterior segment of the mandible, generally appearing as a rather poorly circumscribed lesion, somewhere between the first premolar and central incisor area. This may be caused by impingement of sublingual gland.



Fig. 26-6: Stafne's bone cyst presented as radiolucency below the mandibular canal and surrounded by well corticated border (Courtesy Dr Tapasya Karamore).

Diagnosis

- *Clinical diagnosis*—it is not possible or suspect this disease clinically.
- *Radiological diagnosis*—round radiolucency located below the mandibular canal in molar area will diagnose this condition.

Differential Diagnosis

- *Radicular*—it is associated with carious tooth and radiolucency is attached to the root of teeth.
- *Residual*—pre-extraction radiograph shows tooth with evidence of deep caries.

Management

No treatment is necessary as the patient is asymptomatic.

Functional Disorders of Salivary Gland

Sialorrhea (Ptyalism)

An increased salivary secretion is termed as 'sialorrhea' or 'ptyalism'.

Mechanisms

The secretory innervations of the salivary glands are primarily under the control of the parasympathetic nervous system. Stimulation of the parasympathetic system causes profuse secretion of watery saliva. Some persons are unable to swallow their saliva fast enough to prevent drooling.

Etiology

- *Drugs*—certain drugs have the ability to stimulate salivary flow. Such drugs are known as 'sialogogues'. Increased salivation due to drugs can result from a variety of its pharmacologic effects. Besides acting on the parasympathetic and sympathetic receptors, drug action can lead to sialorrhea by direct CNS stimulation or as a result of nasal and oropharyngeal irritation, which leads to afferent stimulation of salivary nuclei. Drug which can cause sialorrhea are lithium and cholinergic agonists.
- *Local factors*—it can be the result of, different types of stomatitis, ANUG, erythema multiforme.
- *Systemic disease*—paralysis, alcoholic neuritis, Parkinson's disease, epilepsy, Down's syndrome, undetermined neuromuscular disorders or following a head injury or stroke.
- *Protective buffering system*—episodic hypersecretion of saliva or 'water brash' may occur as protective buffering system to neutralize stomach acid in individual with gastroesophageal reflux disease.
- *Miscellaneous*—psychic factor, metal poisoning and facial paralysis.

Clinical Features

- *Drooling*—the salivary flow is more in infancy and childhood, but the drooling observed in infants is related to inadequate swallowing rather than excessive production. Drooling or sialorrhea can be a devastating problem for the affected child or adult.
- *Symptoms*—the problem may range from mild embarrassment and discomfort to emotional and physical impairment. The involved person may require numerous clothings and/or bib changes per day.
- *Signs*—he or she may develop cheek scarring, lip chapping or infection from constant exposure to saliva. The soiling of clothes, carpets, furniture, books and people, often results in social rejection, employment difficulties and stigmatization.

Management

- *Treatment in children*—no treatment is recommended in children less than four years of age with only mild or moderate amount of drooling, which may improve spontaneously.
- *Oral motor training*—oral motor training, intended to improve motor skills, is the key non-surgical management modality and all patients (if appropriate) should have a minimum of six months of this type of therapy format, before any surgical management is considered.

- *Biofeedback*—biofeedback utilizes conditioning techniques to train the patients to swallow more frequently. It has been utilized in aware patients with only a modest drooling problem and who have very motivated parents.
- *Removal of local factors*—situational factors that may contribute to drooling should be eliminated if possible, i.e. dental disease, nasal airway obstruction, poor seating and inappropriate medications.
- *Atropine*—the anti-cholinergic drug, atropine sulfate, has been shown to reduce the amount of resting secretion, intraoral accumulation and pharyngeal-laryngeal pooling of saliva in more than 50% of patients.
 - *Mechanism of action*—the drug is a competitive antagonist of muscarinic actions of acetylcholine. It does not prevent the release of acetylcholine, but antagonizes the effect of this neurotransmitter on the effector cells. This action results in drying of the mouth through reduction of salivary gland secretions. Atropine-induced inhibition of salivation occurs within 30 minutes to one hour. Inhibition peaks within two hours after oral administration, but can persist for up to four hours.
 - *Dose*—the usual oral dose for adults is 0.4 mg, every 4 to 6 hours. In children, the suggested dose is 0.01 mg/kg, but generally not exceeding 0.4 mg, every 4 to 6 hours.
 - *Contraindication*—due to potential side effects, atropine sulfate is contraindicated in patients with asthma, glaucoma or synechia (adhesions) between the iris and lens of the eye.
- *Other drugs*—other drugs which are used as anticholinergics are scopolamine (0.4-0.6 mg), methantheline (50-100 mg) and propantheline (15-30 mg).
- *Surgery*—surgery is a primary recommendation in individuals with a cognitive delay and profuse drooling and secondarily in those that have failed to non-surgical therapy for a minimum of six months.
 - *Relocation of duct*—relocation of submandibular and parotid duct posteriorly to the tonsillar fossa. This will reduce salivary flow and drooling.
 - *Bilateral tympanic neurectomy*—sectioning of chorda tympani destroys parasympathetic innervations to the glands.

Xerostomia

It is the subjective clinical condition of less than normal amount of saliva. It is dryness of mouth, which is a clinical manifestation of salivary gland dysfunction.

Etiology

- *Radiation induced*—ionizing radiation to head and neck region for the treatment of cancer results in pronounced changes in the salivary glands located within the primary beam. The degree of damage caused by the radiotherapy is related to dose-time-volume factor. Damage to the acinar cells has been noted with a single 100 rads dose of X-rays. Radiation sensitivity decreases in following order: the parotid gland, submandibular, sublingual to minor glands. Serous acinar cells appear to be more sensitive to radiation, than the mucus cells. As the dose is increased, disorganization and destruction of the acinar cells occur, resulting in their replacement by fibrous or faulty tissues. Both, the stimulated and unstimulated salivary flow rate decreases dramatically with increasing radiotherapy.
- *Pharmacologically induced xerostomia*—there are about 500 drugs which can cause xerostomia. The classes of drugs which cause xerostomia include anticonvulsants, antiemetics, antihistaminics, anti-hypertensives and antispasmodics. The mode of action for decreased salivary flow is generally related to the para-sympathetic activity, usually an antimuscarinic effect. Other actions that can decrease salivation are generally more miscellaneous and include vasoconstriction of salivary glands, changes in fluid and electrolyte balance and changes in acinar or ductal function.
- *Local factors*—local factors like decreased mastication, smoking and mouth breathing can also lead to xerostomia.
- *Developmental*—developmental abnormalities of salivary glands, tumors, autoimmune states and certain diseases which affect afferent or efferent portions of neural transmission reflex are some of the other causes of xerostomia.
- *Systemic alterations resulting in xerostomia*
 - *Nutritional*—certain deficiency states like pernicious anemia, iron deficiency anemia and deficiency of vitamin A and hormones can cause xerostomia.
 - *Fluid loss*—fluid loss associated with hemorrhage, sweating, diarrhea, vomiting.
 - *Diabetes mellitus*—it is associated with xerostomia.
 - *Sjögren syndrome*—xerostomia is also common in Sjögren syndrome.
 - *Other disease*—systemic diseases, which are accompanied by high temperature and dehydration, usually result in diminished salivation. Xerostomia may also be found in HIV infection, sarcoidosis, and graft versus host resistance.

Clinical Features

- *Effect of xerostomia on oral functions*—patient may notice increased thirst, increased uptake of fluid especially while eating. There is also frequent use of means like chewing gums and consumption of sour candy. Patient

also gets difficulty in swallowing, speech and eating dry food. There is also burning and tingling sensations in the mouth. There is also complaint of frequent oral infections, intolerance to dental appliances and abnormal taste in the mouth.

- *Salivary gland enlargement*—painful salivary gland enlargement is also present.
- *Effect of xerostomia on normal functions*—many times, xerostomia is accompanied by hypofunction of other secretory glands. Blurred vision and ocular dryness. Itching, burning and sandy sensation in eye. There is also dryness of pharynx and skin. Itching and burning sensation of vagina.
- *Clinical signs of xerostomia*—dryness of lining oral mucosa. Oral mucosa appears thin, pale and feels dry. Tongue blade may adhere to soft tissues. Tongue may manifest deficiency by atrophy of the papillae, inflammation, fissuring, cracking and denudation. There is also increased incidence of dental caries.
- *Candidiasis*—pseudo membranous and hyperplastic form of candidiasis occurs. The reason for occurrence candidiasis is absence of normal cleansing and antimicrobial activity of the saliva.
- *Residual saliva*—residual saliva which remains is foamy, thick and ropey.

Management

Stimulation of salivary production

- *Local stimulation*—chewing of gums, mints, paraffin and citric acid containing lozenges and rinses. Disadvantages of it are:
 - Effects are short lived.
 - Frequent application can be inconvenient.
 - Citric acid may irritate oral mucosa.
 - Continuous use may contribute to demineralization.
- *Systemic stimulation*
 - *Bromhexine*—it is a mucolytic and mucokinetic agent, capable of inducing thin copious bronchial secretions. Dose—adults (8 mg TDS), children 1-5 years (4 mg BD) and children 5-10 years (4 mg TDS).
 - *Anethole trithione (ANTT)*—it is a directly acting cholinergic agonist which acts by neurostimulation. Dose 1 to 2 tabs (25 mg) TDS.
 - *Pilocarpine*—pilocarpine is a cholinergic parasympathomimetic agent with a broad range of pharmacologic effects. It increases the secretion by exocrine glands and can affect the sweat, salivary, lacrimal, gastric, pancreatic, intestinal glands and mucosal cells of the respiratory tract. The usual dose is 5 mg, TDS. It produces short duration of (3 hours) increased salivary flow, without the accompanying side effects. It should not be used in patients suffering from asthma.

Symptomatic treatment

- *Salivary substitute*—there are number of salivary substitute available for the treatment of xerostomia. Most commonly contain carboxymethylcellulose or hydroxyethylcellulose as lubricants and variety of artificial sweeteners, preservative and chloride or fluoride salts. Disadvantages are:
 - Their regular use is inconvenient to the patient.
 - Most of them are more viscous than the natural saliva.
 - They are expensive.
 - They fail to provide antimicrobial and other protective functions of natural saliva.
- *Composition of artificial saliva*
 - Carboxymethylcellulose—10 gm/l.
 - Sorbitol—30 gm/l.
 - Potassium chloride—1.2 gm/l.
 - Sodium chloride—0.843 gm/l.
 - Magnesium chloride—0.051 gm/l.
 - Calcium chloride—0.146 gm/l.
 - Dipotassium hydrogen phosphate—0.342 gm/l.
- *Oral hygiene product*—patient should use oral hygiene product which include lactoperoxidase, lysozyme, and lactoferrin.
- *Discontinuous of drug*—drug which is causing xerostomia should be discontinued.

Suggestions to the patient having xerostomia

- *Sweet and tart food*—try very sweet or tart foods and beverages such as lemonade; these foods may help to produce more saliva. (Do not try this in sensitive teeth or sore throat.)
- *Sucking of sugar free candy*—suck on sugar-free hard candy (avoiding those with citric acid), popsicles or chew sugar-free gum. These can help produce more saliva.
- *Sucking ice cubes*—try sucking ice cubes or ice lollies. Home-made lollies can be easily made by freezing fresh juice in ice-cube trays or in special lolly containers with sticks, which can be bought from many kitchenware shops.
- *Don't take following thing*—avoid chewable vitamin C and acidic, sugared lozenges. Avoid dry foods such as cookies, toast and crackers, or soften them with liquids before eating. Avoid chocolates, peanut butter and pastry: they stick to the roof of mouth. Avoid over salty foods.
- *Soft and liquid food*—use soft and liquid foods, which may be easier to swallow.
- *Drink frequently*—have a sip of water every few minutes to help in swallowing and talking more easily. Carry a small water bottle for frequent sips during the day.

Obstructive Disorders

Sialolithiasis

It is the formation of calcific concretions within the parenchyma or ductal system of the major or minor salivary glands. It is also called as 'salivary gland stone' or 'salivary gland calculus'. These are stones within major and minor salivary glands. These are the most common calcifications found in soft tissues of oro-orbital region.

Composition

- *Calculus*—the calculus consists of laminated layers of organic material, covered with concentric shells of calcified material.
- *Crystalline structure*—the crystalline structure is chiefly hydroxyapatite and contains octacalcium phosphate.
- *Chemical composition*—the chemical composition is principally calcium phosphate and carbon with traces of magnesium, potassium, chloride and ammonium.

Etiopathogenesis

- *Neurohumoral mechanism*—a neurohumoral condition, leading to salivary stagnation, results in a nidus and matrix formation.
- *Metabolic mechanism*—in the presence of coexisting inflammation, a metabolic mechanism favors precipitation of salivary salts into the matrix.

Prevalence

Submandibular (83%) calculi are more commonly seen than the parotid (10%) or sublingual (7%) calculi, due to following factors.

- *Anatomic factors*
 - The length and irregular course of Wharton's duct.
 - The submandibular gland and ductal system lies in a dependent position.
 - The greater size and position of the orifice.
 - The orifice is much smaller than duct lumen.
- *Physiochemical factors*
 - High mucin content of saliva.
 - Great degree of alkalinity with high percentage of organic matter.
 - Greater concentration of calcium and phosphate salts.
 - Low content of carbon dioxide.
 - Richness in phosphatase enzyme.

Types

- *Ductal sialoliths*—it is located in the duct of gland.
- *Glandular sialoliths*.

Clinical Features

- *Age and sex*—they are usually encountered in middle aged patients with slight predilection for occurrence in men.
- *Symptoms*—the symptoms of sialolithiasis vary but intra-glandular stones seem to cause less severe symptoms than the extraglandular or intraductal types. On occasions, there may be complete absence of subjective symptoms.
 - *Pain*—many patients complain of moderately severe pain. The occlusion of the duct prevents the free flow of saliva and this stagnation or accumulation of saliva, when under pressure, produces pain.
 - *Swelling*—patient also complains of intermittent transient swelling during meals, which resolves after meals. As the calculus itself rarely blocks a duct completely, the swelling subsides as salivary demand diminishes and as saliva seeps past the partial obstruction.
 - *Systemic symptoms*—if no treatment is instituted, it appears as a pronounced exacerbation characterized by an acute suppurative process with attendant systemic manifestations such as fever and malaise.
- *Signs*
 - *Pus*—pus may exude from the duct orifice.
 - *Surrounding tissue*—the soft tissues surrounding the duct show a severe inflammatory reaction. It appears as swelling, redness and tenderness.
 - *Palpation*—stones in the more peripheral portion of the duct may often be palpated, if they are of sufficient size.
 - *Ulceration*—sometimes, the overlying mucosa may ulcerate over the stone allowing the calculus extend into the oral floor.
 - *Absence of saliva*—no saliva is seen to be coming out through the duct orifice.
- *Swab test*—if stone is present in one duct only then saliva will not come out from that duct. It can be tested by placing two dry swabs one on each orifice and some lemon juice is dropped on the dorsum of the tongue. A minute later patient is asked to move the tongue up. The swab on the orifice of the duct where the stone is impacted will remain dry, whereas the other swab will be wet.
- *Size*—it usually occurs as a solitary concretion varying in size from a few millimeters up to several centimeters.
- *Stones in minor salivary glands*—sialolithiasis of minor salivary gland is a rare occurrence. The most common site is buccal mucosa either near the commissure or in proximity to the mandibular mucobuccal fold. It is more common after the age of 39 years. The lesions appear as firm, freely movable masses, deeply situated into the mucosal surface.

Radiographic Features

- *View taken*—projection for submandibular duct stone is standard mandibular occlusal view and for parotid gland, periapical view in the buccal vestibule. Reduce the exposure to avoid burnout of sialoliths.
- *Sialography*—sialography is indicated when sialoliths are radiolucent. There is ductal dilatation caused by associated sialodochitis. The film usually shows contrast medium present behind the stone.
- *Site*—there is white or gray opacity situated somewhere in the region of the glandular apparatus. It may be solitary or multiple.
- *Radiodensity*—they are almost radiopaque, so that even very small ones are visible in well prepared radiograph (Fig. 26-7).

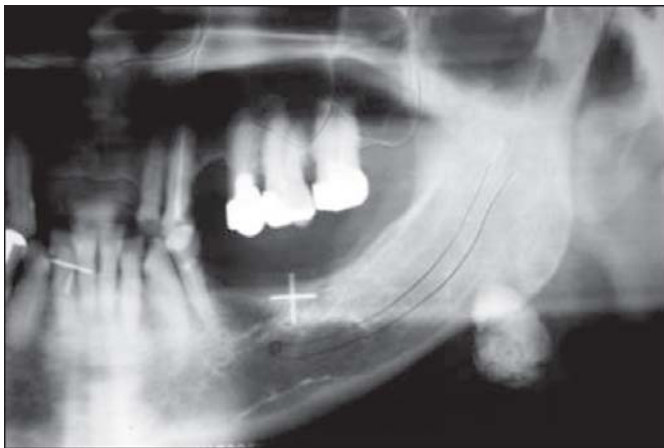


Fig. 26-7: Well defined radiopacity seen at the angle of mandible due to salivary stone (Courtesy Enzo Rovigutti)

- *Shape*—it is usually oval shaped and is cylindrical with multiple layers of calcification.
- *Borders*—smooth borders with even radiodensity.
- *Size*—the size varies from little more than a pinhead up to a length of an inch (Fig. 26-8) or more, with a girth of about 5 mm.
- *CT*—it will show dense radiopaque area in contrast study (Fig. 26-9).

Diagnosis

- *Palpation*—palpation is an indispensable tool in the diagnosis of sialoliths. Palpation of the suspected gland frequently reveals it to be larger or firmer than the normal gland of the opposite side. Digital manipulation will produce a flow of saliva through the duct orifice and will allow visual inspection of the salivary fluid. During examination, the soft tissues overlying the duct should be manually stretched. Often, the physical distortion caused by the presence of calculus will become apparent. In addition yellowish color of the calcific deposits may



Fig. 26-8: Small size salivary stone present as radiopacity in ramus of mandible (Courtesy Enzo Rovigutti).



Fig. 26-9: Well defined radiopaque area seen in submandibular gland (Courtesy Dr Iswar).

be seen through the distended and thinned mucus membrane.

- *Metallic duct probe*—a metallic duct probe can also be valuable. Careful probing of the duct with a metallic probe will indicate the existence as well as the location of calculus.
- *Radiographic examination*—radiographic examination usually reveals the presence of calcific deposits.
- *Sialography*—sialography is an invaluable aid in isolating the sialoliths which had not been identified on the standard intraoral and extraoral radiography.

Differential Diagnosis

- *Gas bubble*—it should not be confused with gas bubbles introduced during sialography. Gas bubbles are more easily removed and are more circular than sialoliths.
- *Hyoid bone*—bilateral on panoramic radiography.
- *Myositis ossificans*—restriction of mandibular movements.
- *Phleboliths*—no sialadenitis present. They are more or less rounded and contain laminations or central dark area.
- *Calcific submandibular lymph nodes*—if painful swelling accompanies the calcified mass, it is usually sialolith.
- *Chondrodystrophia calcificans congenita*—it is sometimes associated with calcification in the neck, which can resemble the submaxillary calculi in the radiographs.

Management

- *Manual manipulation*—in case of small stone, gentle massage of the gland should be done. It will help to move the stone toward the duct orifice. Sialogogues, moist heat and increased fluid intake will also promote the passage of stone.
- *Stone in the submandibular duct*—if the stone is palpated near the orifice of the duct it can be removed by an incision made directly over it through the mucus membrane of the mouth.
- *Stone in the submandibular gland*—in this case, excision of gland is advised.
- *Antibiotics*—if acute infection is present, then antibiotics should be given.
- *Salivary gland endoscopy*—this is newer method which is useful in removal of sialoliths.
- *Lithotripsy*—this is fragmentation of stone in the gland. Nowadays, extracorporeal shock wave lithotripsy has been successful in many patients.

Mucus Plugs

These are incompletely mineralized sialoliths. Clinical symptoms and signs are same as that of stones. The diagnosis of mucus plug is based upon clinical history, radiography and sialograms. If sialograms confirm the intraductal obstructive disease secondary to non-mineralized intraductal object, the diagnosis of mucus plug can be made.

Strictures and Stenosis

These are the rare conditions occurring in the salivary gland area.

Etiology

- *Irritation*—irritation from prosthetic appliances, maloccluded or malpositioned teeth.

- *Acute trauma*—acute trauma with resultant edema and/or scarring.
- *Tumor*—intraductal tumor formation.

Types

- *Papillary obstruction*—it may be either acute ulcerative obstruction or chronic fibrotic stenosis. Acute ulcerative obstruction is usually caused by acute trauma to the papilla and is treated conservatively with saline rinses and salivary gland massage. The ulcer generally heals without scarring and the symptoms will subside in such cases. In chronic fibrotic papillary obstruction irritations to papilla has been recurrent and scarring exists.
- *Duct obstruction*—it may be due to a variety of factors. In cases of ductal obstruction secondary to acute trauma treatment is directed towards providing the duct patency until the edema is resolved. When the ductal obstruction occurs secondary to irritation or scar contracture, sialograms are helpful in localizing the status of gland. If the gland is healthy, progressive and frequent dilatation of involved duct with lacrimal probes is generally successful in relieving the symptoms and signs. If this does not prove beneficial, ductoplasty is indicated.

Foreign Bodies

Rarely, foreign bodies become lodged within Wharton's duct and less commonly in Stensen's duct. Toothbrush bristles, toothpicks, spikes of wheat, fish bone, portions of fingernail have been reported within the salivary gland duct and act as causes of obstructive and/or inflammatory disease of the salivary gland.

Extraductal Causes

Muscle pressure, tumors, enlarged lymph nodes and denture flanges associated with the primary salivary duct can cause obstructive signs and symptoms.

Parotid Fistula

It may arise from parotid gland or duct. It may be internal, when it opens inside the mouth and external, when it opens to the exterior.

Causes

- *Traumatic*—penetrating injury particularly by glass splinters.
- *Parotid abscess*—rupture of parotid abscess.
- *Inadvertent incision*—inadvertent incision while draining of parotid abscess.
- *Complication*—complication of superficial parotidectomy.

Clinical Features

- *Symptoms*—the main complaint is opening in the cheek with discharge.
- *Signs*—discharge come out only during meals.
- *Adjacent skin*—there may be excoriation of the neighborhood skin.

Sialograms

- It will give an indication whether the fistula is in relation to the main duct, ductile or to the gland.

Diagnosis

- *Clinical diagnosis*—discharged from Stensen's duct after trauma or surgery.
- *Radiological diagnosis*—it will exactly diagnose the parotid fistula.

Management

- *Reconstruction of the duct*—it is done by surgical approach.

Cysts of Salivary Gland

Mucocele

It is a term used to describe the swelling caused by pooling of saliva at the site of injured minor salivary gland. It is also called as 'mucus extravasation phenomenon, mucus escape reaction'. It is not true cyst it lacks an epithelial lining.

Etiopathogenesis

- *Trauma*—it is caused by laceration of a minor salivary gland duct by trauma resulting in extravasation of mucus into the connective tissue. There is accumulation of mucus in the connective tissue and with the continuous pooling of saliva a clearly demarcated cavity develops which has no epithelial lining.

Clinical Features

- *Age and sex*—mucus extravasation cysts occur in younger patients. The reason for occurrence in young people as they are more prone to trauma that induce mucin spillage. It is equal in both sexes.
- *Site*—they are very common and occur most frequently on the inner aspect of lower lip (**Fig. 26-10**); but may also occur on the palate, cheek, tongue and floor of mouth. The lesion may lie fairly deep in the tissues or may be exceptionally superficial.
- *Symptoms*—patient may complain of painless swelling which is frequently recurrent. The swelling may suddenly develop at meal time and may drain simultaneously at intervals.



Fig. 26-10: Mucus extravasation cyst seen on lower lip (Courtesy Dr Parate).

- *Size*—the mucocele may be only 1-2 mm in diameter, but is usually larger; majority of them being between 5 and 10 mm in diameter.
- *Appearance*—superficial cyst appears as bluish mass, as the thin overlying mucosa permits the pool of mucus fluid to absorb most of the visible wavelength of light (**Fig. 26-11**). If inflamed, it is fluctuant, soft, nodular and dome shaped elevation. Deeper lesions have the color of normal mucosa and are firmer.

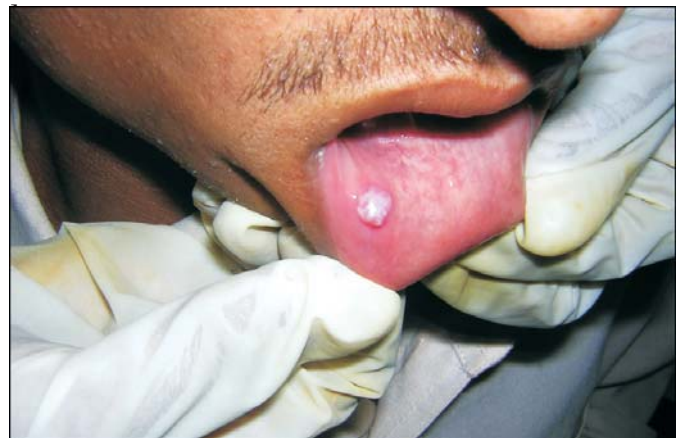


Fig. 26-11: Bluish color of mucocele seen on lower lip.

- *Shape*—the swelling is round or oval or dome shaped (**Fig. 26-12**).
- *Consistency*—it is either soft or hard depending upon the tension in the fluid. It cannot be emptied by digital pressure.
- *Aspiration*—on aspiration, it yields sticky viscous clear fluid.
- *Superficial mucocele*—it is present as single or multiple tense vesicles that measures 1 to 4 mm in diameter. The lesion burst leaving shallow painful ulcer that heals



Fig. 26-12: Dome-shaped swelling of mucocele (Courtesy Dr Parate).

within a few days. It is seen on soft palate and posterior buccal mucosa.

- **Recurrence**—patient may give history of recurrent swelling which ruptures and reappears again after some time.

Diagnosis

- **Clinical diagnosis**—dome shaped soft swelling on lower lip which is lateral to midline is typical features of mucocele.
- **Laboratory diagnosis**—in biopsy, it shows vacuolated macrophages which are sometimes called as ‘muciphage’.

Differential Diagnosis

- **Vascular lesion and superficial non-keratin cyst**—aspiration should be done.
- **Early Mucoepidermoid tumor and adenocarcinoma**—induration is present.

Management

- **Surgical excision**—complete excision of the mucocele should be done under local anesthesia. To avoid recurrence, adjacent minor salivary gland should also be removed which are feeding the lesion.
- **Cryosurgery**—surgery with cryoprobe is also helpful in managing the mucocele.

Ranula

It is derived from Latin word ‘*Rana tigrina*’, i.e. frog belly. The term ranula is used for the mucoceles occurring in the floor of the mouth, in association with ducts of submandibular or sublingual glands.

Types

- **Superficial**—the superficial variety may develop as a retention or extravasation phenomenon associated with trauma to one or more of the numerous excretory ducts of the sublingual salivary gland.
- **Plunging or cervical**—it ramifies deeply into the neck.

Clinical Features

- **Age and sex**—it is usually seen in children and young adults with no sex predilection.
- **Site**—the typical position is on the floor of the mouth (Fig. 26-13), below the tongue and on the side of frenum. They are usually unilateral.



Fig. 26-13: Unilateral swelling seen on left side in floor of mouth (Courtesy Dr Datarkar)

- **Symptoms**—it develops as slowly enlarging painless mass on one side of the floor of mouth. When the swelling suddenly grows, it may be painful. Big ranula may cause difficulty in speech or eating.
- **Appearance**—they produce blue shaped swelling (Fig. 26-14) like a frog’s belly, hence it was given the term ‘ranula’ (‘ranula’ in Greek mean frog’s belly). The overlying mucosa of the swelling is normal in appearance.
- **Shape**—it is spherical or dome shaped with only superficial half is visible.
- **Size**—it is smaller in early morning and largest just before meals, due to increased secretory activity in periods of gustatory stimulation and water absorption from the pooled mucus during inactive period.
- **Consistency**—it is soft and tends to fluctuant. It cannot empty by pressure and is non-pulsatile.
- **Fluctuation and transillumination**—both tests are positive. The ranula is typically known as brilliant translucent swelling.
- **Aspiration**—aspiration yields sticky clear fluid.
- **Plunging ranula**—when intrabuccal ranula has a cervical prolongation, it is called as deep or plunging ranula.



Fig. 26-14: Blue shaped swelling seen of ranula.

- *Origin*—it is derived from cervical sinus.
- *Location*—it lies along the posterior border of the mylohyoid muscle and appears in the submandibular region.
- *Appearance*—sometimes, plunging ranula herniates through the mylohyoid muscle and cause a swelling in suprahyoid or infrahyoid region.
- *Bidigital palpation*—to palpate plunging ranula, bidigital palpation should be performed. One finger is placed inside the mouth on the ranula and the other finger is placed on the swelling in the submandibular region. If pressure on the first finger causes sense of fluctuation on 2nd finger or vice versa, then it is plunging ranula.

Radiological Features

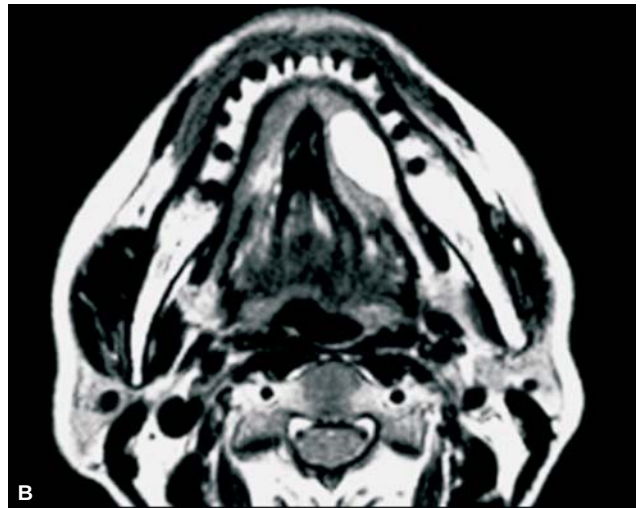
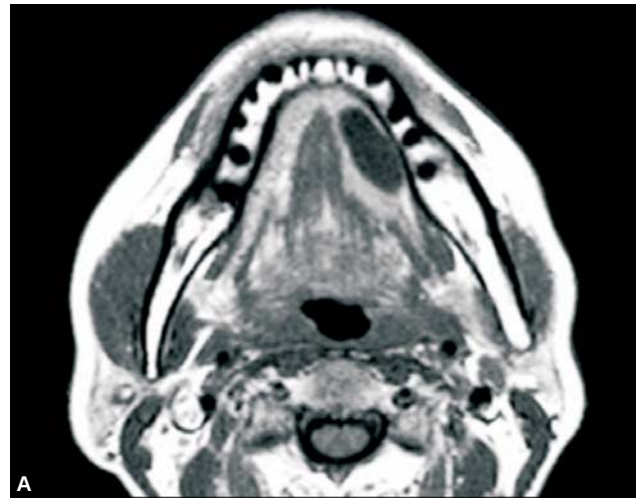
- *CT features*—On CT, the lesion is observed as a homogeneous, water density mass with clear boundary (Fig. 26-15A).
- *MRI features*—on MR images, the lesion shows low signal intensity on T1 weighted images; very high signal intensity on T2 weighted images. On sonograms, the plunging ranulas are observed as a well-delineated, but not round, anechoic mass (Fig. 26-15B).

Diagnosis

- *Clinical diagnosis*—blue shaped swelling in floor of mouth in lateral position will go in favor of ranula.
- *Radiological diagnosis*—MRI and CT scan will demonstrate lesion.

Differential Diagnosis

- *Sublingual dermoid*—it is more often in midline and it is not translucent, while ranula is translucent.
- *Submandibular lymph nodes swelling*—it is hard or firm in consistency.



Figs 26-15A and B: Simple ranula. (A) T1 weighted MR image shows lower signal intensity than that of muscles in the left sublingual region; (B) T2 weighted MR image of the same case. Note the remarkably high signal intensity of the lesion (Courtesy M Shimzu).

Management

- *Surgical excision*—they are best treated by surgical excision including a portion of the surrounding tissues.
- *Partial excision with marsupialization*—the major part of the cyst wall together with its overlying mucus membrane is excised.

Salivary Duct Cyst

It is also called as '*mucus retention cyst, mucus duct cyst, and sialocyst*'. It is true cyst as it is lined by epithelium.

Etiopathogenesis

- *Obstruction*—it is caused by obstruction of minor salivary gland duct which causes the backup of saliva. This continuous pressure dilates the duct and forms a cyst like lesion.

Clinical Features

- *Age and sex distribution*—retention cysts occur most often in older patients with no predilection for any sex.
- *Site*—this is most common in parotid gland. Intraorally, it is common on floor of mouth, buccal mucosa and lips.
- *Appearance*—it is slow growing and looks like mucoceles. It is soft, fluctuant swelling which appear bluish.
- *Multiple retention cyst*—in some cases, patient may develop multiple painful nodules which demonstrated as dilated ductal orifices on the mucosal surface.

Diagnosis

- *Clinical diagnosis*—it is very difficult to differentiate between mucocele and retention cyst.
- *Laboratory diagnosis*—biopsy shows lining of epithelium which consists of cuboidal, columnar or atrophic squamous epithelium surrounding thin or mucoid secretion in the lumen.

Management

- *Surgical excision*—it is treated by conservative surgical excision.
- *Antibiotics*—in some cases of multiple retention cyst, antibiotics like erythromycin and chlorhexidine mouth is helpful in relieving the pain of the patient.
- *Sialogogues*—this is helpful in stimulating salivary flow and thereby preventing an accumulation of mucin.

Asymptomatic Enlargement of the Salivary Gland

Sialosis (Sialadenosis)

It is characterized by non-neoplastic non-inflammatory enlargement of the salivary gland.

Etiopathogenesis

- *Systemic disease*—the condition is found in association with systemic diseases especially cirrhosis, diabetes, ovarian and thyroid insufficiency, alcoholism, general malnutrition, anorexia nervosa, and malnutrition.
- *Neurogenic medication*—neurogenic medication like antihypertensive drugs, psychotropic drugs and sympathomimetic drugs can cause sialosis.
- *Mechanism*—above disease may result in dysregulation of the autonomic innervations of the salivary acini causing an aberrant intercellular secretory cycle.

Clinical Features

- *Age and sex distribution*—it more commonly affects the females. As such there is no age predilection.

- *Site*—the enlargement is usually bilateral and may present a course of recurrent painless enlargement of gland. The parotid gland is more frequently affected.
- *Appearance*—swelling of the preauricular portion of the parotid gland is the most common symptom, but retromandibular portion of the gland may also be affected.

Radiological Features

- *Sialography*—on sialography, leafless tree appearance is seen (**Fig. 26-16**). This appearance is caused by compression of finer duct by hypertrophic aciner cells.

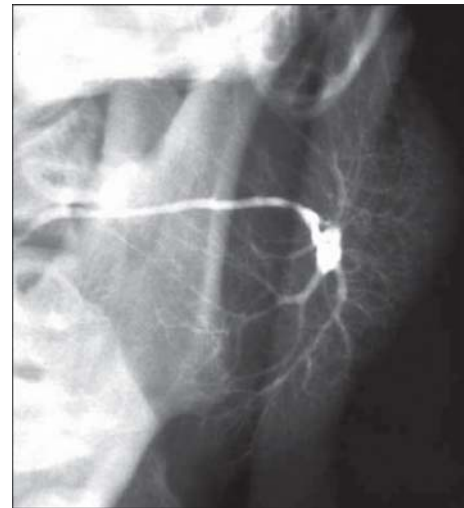


Fig. 26-16: Leafless tree appearance seen in sialosis (Courtesy M Shimzu)

Diagnosis

- *Clinical diagnosis*—bilateral non-inflammatory enlargement of parotid gland gives one clue to the diagnosis.
- *Radiological diagnosis*—leafless tree appearance seen in this disease.
- *Laboratory diagnosis*—a characteristic alteration in the chemical constituents of saliva is a distinguishing feature of sialosis. Significant elevation of salivary potassium and concomitant decrease in salivary sodium is observed.

Management

- *Control of underlying cause*—this is most important management for sialosis.
- *Partial parotidectomy*—it can be performed if swelling becomes cosmetic concern.

Allergic Sialadenitis

In some cases, it may not appear as a true hypersensitivity reaction but rather as a toxic or idiosyncratic reaction to drugs that cause decreased salivary flow, resulting in secondary infection.

Etiopathogenesis

- **Drugs**—various drugs which have been reported to cause allergic sialadenitis include sulfisoxazole, phenothiazines, iodine containing compounds, mercury, thiouracil and phenylbutazone.
- **Mechanism**—the exact mechanism of salivary gland enlargement and loss of function following administration of these drugs is not known. Most of the drugs cause decrease in capillary permeability, whereas others cause sodium and chloride retention, which subsequently leads to edema.

Clinical Features

- **Appearance**—the clinical appearance of allergic sialadenitis varies, but in most of the cases, there is bilateral parotid gland enlargement following the administration of the drug.
- **Symptoms**—the enlargement may be painful and is usually associated with conjunctivitis and skin rashes.

Diagnosis

- **Clinical diagnosis**—enlargement of gland with skin rashes and conjunctivitis.

Management

- It is a self-limiting disease and needs no treatment. But in some cases, secondary bacterial infection may develop and need treatment.

Associated with Malnutrition or Alcoholism

Asymptomatic enlargement of the parotid gland may occur in patients with nutritional deficiency, especially vitamin A and alcoholism. The patient usually gives history of long-term gradually increasing swelling of the parotid gland.

Clinically, the swelling is asymptomatic and can occur bilaterally. Palpation of enlarged gland reveals a normal tone and non-tender swelling. Simultaneous swelling of the submandibular salivary gland is a rare finding. The flow has been reported to be increased, but composition of the saliva remains unchanged, except for the elevated amylase level.

Viral Infection

Various viruses like paramyxovirus, cytomegalovirus, parainfluenza type-3 and coxsackie virus may infect salivary glands and cause its enlargement.

Mumps

It is also called as 'epidemic parotitis'. It is an acute contagious viral infection, characterized chiefly by unilateral or

bilateral swelling of the salivary glands. It mainly affects major salivary glands, but also affects the testis, meninges, pancreas, heart and mammary glands. It is caused by paramyxovirus. Incidence of mumps decreases after vaccine is available since 1967.

Transmission

- **Saliva and urine**—it usually spreads from human reservoir, by airborne infection of infected saliva and possibly urine.

Clinical Features

- **Age and sex**—it is more common in boys than in girls and most often seen between the age of 5 and 15 years.
- **Incubation period**—it is of 2 to 3 weeks and patient is contagious from one day before clinical appearance of the lesion.
- **Site**—the parotid gland is most commonly involved and it is usually bilateral. Submandibular gland may also be involved, although this is less noticeable and cause less pain. Both the parotid glands may involve simultaneously, but more commonly one parotid gland swells 24 to 48 hours after the other.
- **Prodromal symptoms**—it is preceded by onset of headache, chills, moderate fever, vomiting and pain below the ear which lasts for about one week.
- **Onset**—it is then followed by sudden onset of salivary gland swelling which is firm somewhat rubbery or elastic and without purulent discharge from the salivary gland duct.
- **Symptoms**—it produces pain upon mastication especially while eating sour food.
- **Signs**—the enlargement of parotid gland causes elevation of ear lobule. Swelling may extend upto posterior border of mandible.
- **Sublingual gland involvement**—its involvement of sublingual gland occur bilaterally. It may produce swelling in the floor of mouth.
- **Ductal papilla**—papilla on the opening of parotid duct is often puffy and reddened.
- **Course**—most of the cases are self-limiting, with salivary gland enlargement subsiding within a week.

Complications

- **Orchitis**—When mumps occur in adult males, orchitis (inflammation of the testis) is of great danger and ensues in 20% of the cases. It may result in sterility.
- **Pancreatitis**—involvement of pancreas producing acute pancreatitis often causes an elevation in serum lipase.
- **Meningitis**—meningitis and encephalitis can occur as complication of the disease.

- *Others*—deafness, mastoiditis, meningoencephalitis, epididymitis and myocarditis, have also been reported.

Diagnosis

- *Clinical diagnosis*—the presence of parotitis and accompanying systemic signs of viral infections.
- *Laboratory diagnosis*—salivary amylase level is increased. A paramyxovirus may be isolated from saliva for as long as 6 days before and up to 99 days after the appearance of salivary gland swelling.

Management

- *Vaccination*—prevention with live attenuated vaccine is the best method of controlling the disease. Vaccine should be given in 12 to 15 months of life. It should be repeated at the age of 4 to 5 years.
- *Symptomatic treatment*—symptomatic treatment is given to control pain and swelling. Mainly non-aspirin analgesic and antipyretic should be given.
- *Rest*—bedrest is recommended to minimize the chances of orchitis.
- *Diet restriction*—patient should avoid sour foods and drinks to decrease salivary gland discomfort.

Cytomegalovirus Inclusion Disease

It is also called as 'salivary gland virus disease'. It is caused by cytomegalovirus, a herpes virus. It is common in immunosuppressed adult. Although it is congenital in nature, it is usually secondary to concurrent disease which has caused debilitation.

It affects primarily in newborn infants and children, but adults are also affected. In newborns, infection is generalized and is usually fatal with involvement of liver, lungs and central nervous system. Infants who survive the infection may have permanent central nervous system involvement, including mental retardation and seizures. There may be hepatosplenomegaly, hemolytic anemia and hemorrhagic tendency. It may cause clinical disease of salivary gland, causing enlargement of the gland.

Bacterial Infection

Bacterial infection of salivary gland may be recurrent and generally develops owing to spread of microorganisms from the oral flora along the excretory duct.

Acute Bacterial Sialadenitis

It is also called as 'acute suppurative parotitis'. Most of the bacterial infection occurs as results of ductal obstruction of decreased salivary flow.

Etiology

- *Microorganisms*—it is most commonly caused by penicillin resistant *Staphylococcus aureus* or streptococci viridians.
- *Host factor*—it may be caused due to decreased host resistance, decreased salivary secretion and decreased bactericidal effect of saliva.
- *Predisposing factors*—it can occur in conditions such as dehydration, malnutrition, cancer and surgical infections.
- *Surgical procedure*—it is common when major surgical procedure is carried out in patient with poor oral hygiene.
- *Oral hygiene*—poor oral hygiene is an important contributory factor.
- *Drugs*—drugs like anti-Parkinson's, diuretics and antihistaminic have been reported to be a contributory factor for acute bacterial sialadenitis.

Clinical Features

- *Age*—most of the cases occur in adults but neonates and childhood form of the disease may occur.
- *Site*—unilateral involvement of parotid gland is common.
- *Prodromal symptoms*—it begins with the elevation of body temperature and sudden onset of pain at the angle of the jaw which is intense when the extensive infection is contained within the confines of the parotid capsule.
- *Symptoms*—the localized symptoms are accompanied by fever, leukocytosis and other generalized signs and symptoms of acute bacterial infection.
- *Signs*—parotid gland is tender, enlarged and the overlying skin is warm and red. The swelling usually causes elevation of the ear lobule (**Fig. 26-17**) and the overlying skin is characteristically warm and red.



Fig. 26-17: Enlargement of parotid gland with elevation of ear lobules in acute bacterial sialadenitis.

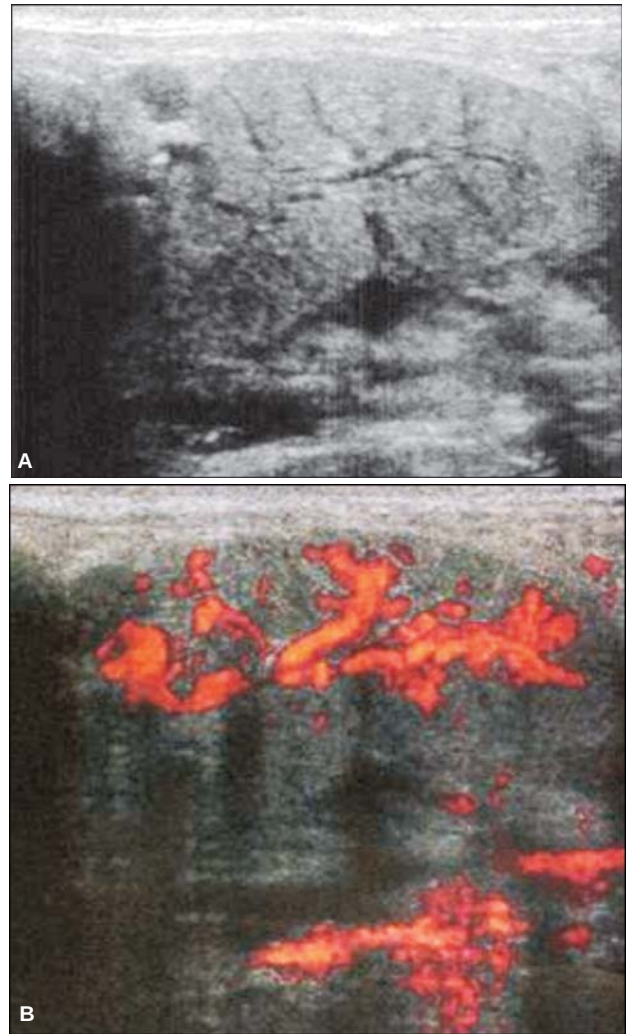
- *Salivary duct*—early in the disease, flecks of purulent material can be expressed from the salivary duct orifice. Intraorally, the parotid papilla may be inflamed and pus may exude or be milked from the duct of the affected gland.
- *Lymph nodes*—cervical lymphadenopathy usually develops.
- *Spread of infection*—if infection is not eradicated, pus may penetrate the gland and spread into the surrounding tissues by following routes:
 - Downwards into the deep facial plane of neck.
 - Backwards into the external auditory canal.
 - Outwards into the skin of face.
- *Postoperative parotitis*—it develops 5 to 7 days after the operation.
 - *Symptoms*—the clinical symptoms are sudden in onset with development of warm, firm or indurated swelling at the angle of jaw and over the cheek. It may be associated with local pain and tenderness. The swelling may cause trismus.
 - *Signs*—the swelling is usually brawny edematous and often causes elevation of ear lobule. The pus can be observed flowing from the opening of duct or it can be produced by a gentle milking action of cheeks.
 - *Prevention*—certain preventive measures should be taken to prevent such complication. The fluid and electrolyte balance of the body should be maintained during postoperative period. Good oral hygiene should be established prior to and after the surgery.
- *Subacute necrotizing sialadenitis*—it is salivary inflammation occurs in teenager and young adults. It involves minor salivary gland. It presents as painful nodules which is covered by intact erythematous mucosa.

Radiological Features

- *Sonography*—the affected gland shows swelling, increased vascularity, however, echo level is not decreased.
- *CT features*—when the glands are involved in cellulitis, swelling of the glands and obscuration of the glands' contour can be observed. On enhanced CT, the affected glands are seen with a higher CT values compared with the normal side because of the increased vascularity (**Figs 26-18A and B**).

Diagnosis

- *Clinical diagnosis*—tender parotid gland with elevation of ear lobule with purulent material seen from Stensen's duct.
- *Radiological diagnosis*—sonography and computed tomography will diagnose this condition.



Figs 26-18A and B: Sonograms of acute sialadenitis of the submandibular gland scanned perpendicular to the mandibular plane (left side: cranial, right side: caudal). (A) The left submandibular gland shows swelling, however, normal echo level. The black lines inside the gland are not dilated ducts, they are blood vessels; (B) The gland shows increased vascularity (Courtesy Dr M Shimizu).

Management

- *Oral hygiene*—meticulous oral hygiene should be practiced. Oral hygiene should be maintained by debridement and irrigation.
- *Diet*—soft diet should be given as chewing is painful.
- *Antibiotics*—it is treated aggressively with antibiotics. Even death can result in debilitated patients. Specimen of purulent material should be immediately sent to laboratory for sensitivity and culture. Treatment usually starts with high dose of parenteral antibiotics active against penicillin resistant *Staphylococcus*.
- *Electrolyte balance*—the patient must be adequately hydrated and the electrolyte balance should be properly maintained with intravenous fluids.
- *Stimulation of saliva*—salivation should be stimulated to facilitate drainage by sucking the sour hard candy.

- **Surgical drainage**—if improvement does not occur, surgical drainage of the affected gland should be performed.

Chronic Bacterial Sialadenitis

It is usually caused by *Streptococcus viridans*, *E coli* or *proteus*. As compared to acute parotitis, it can be seen in normal children or in adults.

Etiology

- **Ductal obstruction**—it results due to recurrent or persistent ductal obstruction.
- **Other causes**—congenital stenosis, Sjögren's syndrome, or previous viral infection or allergy.

Clinical Features

- **Age**—the childhood form commonly begins between the ages of 3 and 5 years and is usually unilateral.
- **Symptoms**—the pain is usually minimal and antibiotic therapy resolves the infection within a week.
- **Appearance**—it appears as a unilateral swelling at the angle of the jaw in a patient with the history of similar occurrence (**Fig. 26-19**).
- **Signs**—salivary flow is accompanied by flecks of purulent material. After several recurrences, fibrosis of the glandular parenchyma occurs, which leads to decreased salivary flow.



Fig. 26-19: Chronic bacterial sialadenitis presenting as swelling at angle of jaw (Courtesy Dr Chole).

Radiological Finding

- Sialograms show multiple ectasias and dilatation of the main excretory duct (**Fig. 26-20**).

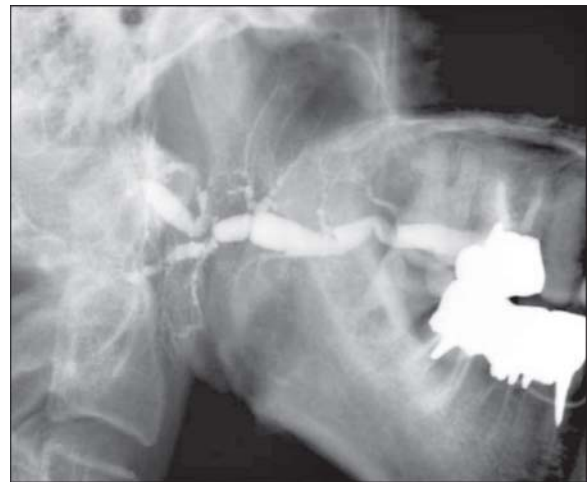


Fig. 26-20: Chronic sialadenitis showing dilatation of excretory duct and multiple ectasia (Courtesy M Shimizu).

Diagnosis

- **Clinical diagnosis**—minimum pain in parotid gland area with purulent material seen from duct of gland.
- **Radiological diagnosis**—dilation and multiple ectasia seen on the radiograph.

Management

- **Radiation therapy**—it is used extensively to cause fibrosis of salivary gland, but there are then increase incidences of head and neck tumors.
- **Surgical removal**—total removal of parotid gland in cases of intractable cases. But is risk of facial nerve palsy.
- **Antibiotics injection**—intraductal injection of erythromycin or tetracycline. In it, cannulate the duct and anesthetize the area with an infusion of lidocaine directly in the duct system, which is followed by infusion of antibiotics into the duct at a concentration of 15 mg/ml.
- **Ligation of Stensen's duct**—ligation of Stensen's duct is a relatively simple procedure and can lead to fibrosis.

Chronic Sclerosing Sialadenitis

It is chronic inflammatory disease. It is also called as '*Kuttner's disease*' and is common in submandibular gland.

Pathogenesis

- It is caused by salivary ductal calculi causing subsequent pyogenic bacterial infections. It can lead to chronic inflammation, ensuring in the atrophy of mucus and serous cells and hyperplasia of the involved connective tissue leading to formation the tumor like swollen masses.

Clinical Features

- **Appearance**—there is enlargement of the glands, resulting in fibrous tumor like masses.

Radiographic Features

- *Sialectasis*—dots or blobs of contrast medium within the gland: appearance known as sialectasis caused by inflammation of the glandular tissue, producing saccular dilatation of the acini.
- *Sausage link appearance*—dilated ductal lumen and constricted ductal system (sausage-link appearance).
- *Absence of terminal branches*—in cases of chronic sclerosing sialadenitis there is absence of terminal ductal branches and presence of constricted ductal lumen within the gland.

Diagnosis

- *Clinical diagnosis*—not so specific.
- *Radiological diagnosis*—sialectasis with sausage link appearance with absence with terminal branches will diagnose this condition.

Management

There is no specific treatment for this disease. In some cases removal of gland is recommended.

Autoimmune Disorders

Sjögren's Syndrome

It is a chronic inflammatory disease that predominately affects salivary, lacrimal and other exocrine glands. It was first described by *Henrik Sjögren* in 1933. It predominately affects middle aged and elderly women.

Types

- *Primary Sjögren's syndrome*—it is also called as sicca syndrome and it consists of dry eyes (xerophthalmia) and dry mouth (xerostomia).
- *Secondary Sjögren's syndrome*—it consists of dry eyes, dry mouth and collagen disorders usually rheumatoid arthritis or systemic lupus erythematosus.

Etiology and Pathogenesis

- *Immunological findings*—the lesion in this syndrome is immunologically mediated inflammatory exocrinopathy. It begins with periductal infiltration of the tissue by mononuclear cells.
- *Autoantibodies*—the B cell hyperactivity may result from deficiency of suppressor T lymphocytes or B lymphocytes by producing autoantibodies against them. Antinuclear antibodies found in patients with Sjögren's syndrome are directed against many nuclear antigens, most commonly to DNA histone. Patients with secondary Sjögren's syndrome tend to develop antibodies against

the EBV-associated nuclear antigen antibodies RANA (rheumatoid arthritis nuclear antigen).

- *Polyclonal hyperglobulinemia*—serum immunoglobulin levels of IgG and IgM are also raised.
- *B₂ microglobulin*—serum salivary level of B₂ microglobulin is raised in minority of patients and correlates with salivary lymphocyte infiltrate.
- *Immune complex*—circulating immune complexes are found in patients with primary and secondary Sjögren's syndrome.
- *Cell mediated immune response*—delayed hypersensitivity response to skin testing is more depressed in patients with secondary Sjögren's syndrome than in patients with primary Sjögren's syndrome. Lymphokine production in response to antigen present in normal salivary tissue, is increased in patients with Sjögren's syndrome.
- *Natural killer cell activity*—augmented natural killer cell activity is impaired in some patients with Sjögren's syndrome. Although natural killer cells provide a defense against viral infection and tumor cell, their role in Sjögren's syndrome is unclear.
- *Virologic aspect*—culture of saliva does not show any specific microorganisms and serologic studies have failed to show increased titers of antibodies, except to cytomegalovirus (CMV).
- *Genetic aspect*—genetic effects of Sjögren's syndrome depend on HLA-linked and non HLA-linked genes. Relatives of a patient with Sjögren's syndrome often show a high incidence of connective tissue diseases. Primary Sjögren's syndrome is associated with HGLA-B₈ and DR₃ and secondary Sjögren's syndrome is associated with HLA-B₈ DR₄ and BW₄₄.
- *Lympho-proliferative malignancy*—enlargement of salivary glands in patients with Sjögren's syndrome is occasionally massive and associated with enlargement of regional lymph nodes, a condition known as 'pseudo lymphoma'. Malignant B cell lympho-proliferation has been shown to affect patients with Sjögren's syndrome.

Clinical Features

- *Age and sex distribution*—it is more commonly seen in middle age adults. It is most commonly found in females with 90% of cases are reported in them.
- *Eyes*—the effect on eye is called as keratoconjunctivitis sicca (dry). The patient usually complains of dry eyes or continuous irritation in the eyes. Severe lacrimal gland involvement may lead to corneal ulceration as well as conjunctivitis.
- *Connective tissue disorders*—in patients with secondary Sjögren's syndrome, rheumatoid arthritis is typically long standing and clinically obvious feature. Patients may have small joint and ulnar deviation of fingers and rheumatoid nodules.

- *Dryness of other organ*—dryness of pharynx, larynx and nose are noted by some patients. This is accompanied by lack of secretion in the upper respiratory tract, may lead to pneumonia. Vaginal dryness may be also complained by some females.

Oral Manifestation

- *Symptoms*—xerostomia is a major complaint in most of the patients. But many patients do not complain of dry mouth, but rather of an unpleasant taste, difficulty in eating dry food, soreness or difficulty in controlling dentures.
- *Signs*—Pus may be emitted from the duct. Angular stomatitis and denture stomatitis also occur.
- *Enlargement*—dry mouth may be accompanied by unilateral or bilateral enlargement of parotid gland, which occurs in about one-third of the patients and may be intermittent. Enlargement of submandibular gland may also occur.
- *Saliva*—clinically, the mouth may appear moist in early stages of Sjögren's syndrome but later, there may be lack of the usual pooling of saliva in the floor of the mouth and frothy saliva may form along the lines of contact with oral soft tissue.
- *Mucosa*—in advanced cases, the mucosa is glazed, dry and tends to form fine wrinkles. Soreness and redness of mucosa is usually the result of candidial infection.
- *Speech*—in some patients, there may be 'clicking' quality of their speech, caused by sticking of the tongue to the palate.
- *Tongue*—the tongue typically develops a characteristic lobulated, usually red surface with partial or complete depapillation. There is also decrease in number of taste buds, which leads to an abnormal and impaired sense of taste.
- *Dental caries*—dental caries is severe and gross accumulation of plaque may be obvious.
- *Periodontal disease*—periodontal disease can also occur.
- *Acute bacterial sialadenitis*—Sjögren's syndrome is the most common underlying cause of acute bacterial sialadenitis in ambulated patients. Such infections are usually either staphylococcal or pneumococcal and usually cause swelling of the salivary gland. The overlying skin is red, tender and shiny.
- *Lymph nodes*—the regional lymph nodes may be enlarged and tender.

Radiological Features

- *Sialography*—if the salivary flow rate is equivocal, sialography can be used to detect the damage. The most typical finding in Sjögren's syndrome is that of

'sialectasia', which typically produces a 'snowstorm appearance' (Fig. 26-21) as a result of leakage of contrast medium. Atrophy of ductal tree may also be seen; emptying of the duct is also typically delayed. In some cases it will show 'cheery blossom' or fruit laden branchless tree (Fig. 26-22) appearance of the obstructed ductal system.

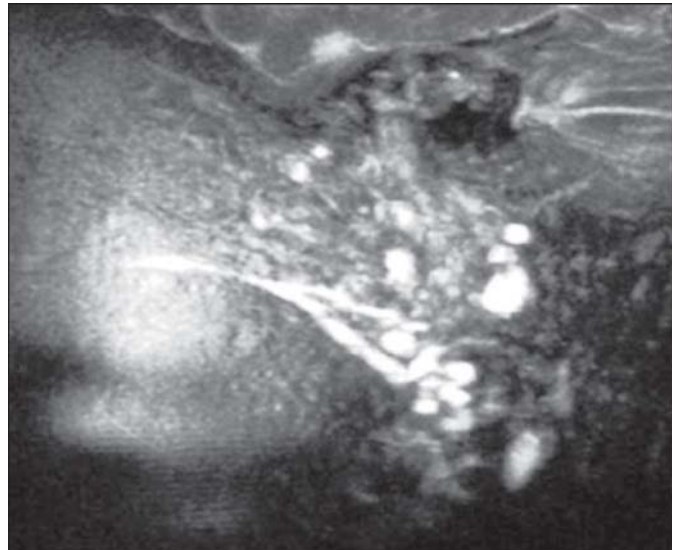


Fig. 26-21: Snowstorm appearance seen in Sjögren's syndrome on sialography (Courtesy M Shimizu).



Fig. 26-22: Branchless fruit laden tree appearance seen in Sjögren's syndrome (Courtesy M Shimizu).

- *Salivary scintiscanning*—salivary scintiscanning with technetium pertechnetate may be useful in demonstrating impaired salivary function in patients with Sjögren's syndrome. The changes correlate with both salivary flow rate and labial gland abnormalities.

Diagnosis

- *Clinical diagnosis*—xerostomia, keratoconjunctivitis sicca with parotid enlargement may aid in diagnosis of Sjögren's syndrome
- *Sialography*—snowstorm and branchless fruit laden tree appearance.
- *San Diego criteria for diagnosis of Sjögren's syndrome.*

Primary Sjögren's syndrome	Secondary Sjögren's syndrome
• Ocular dryness—Schirmer's test less than 8 mm wetting per 5 minute, positive rose Bengal staining of cornea • Dry mouth—decreases parotid flow Lashley cups method • Systemic autoimmunity—elevated Rh factors, elevated antinuclear antibody and presence of anti-SS and anti-SS-B antibodies	• Signs and symptoms of primary Sjögren syndrome • Rheumatoid arthritis, SLE, Polymyositis, scleroderma or biliary cirrhosis • Exclusion of sarcoidosis, pre-existing lymphoma, HIV, Hepatitis B or C, primary fibromyalgia and known cause of keratitis sicca and salivary gland enlargement.

Laboratory Investigations

- *Rose Bengal staining test*—keratoconjunctivitis sicca is characterized by corneal keratotic lesion, which stains pink when 'rose Bengal' dye is used.
- *Schirmer test*—the reduced lacrimal flow rate is measured by this test. A strip of filter paper is placed in between the eye and the eyelid to determine the degree of tears which should be measured in millimeter. When the flow is reduced to less than 5 mm in a 5 minute sample, patient should be considered positive for Sjögren's syndrome.
- *Sialometry*—salivary flow rate estimation is a sensitive indicator of salivary gland function. Parotid glands make the major contribution to total salivary flow and are the most consistently affected glands in patients with Sjögren's syndrome. Stimulated flow rate in symptomatic primary and secondary Sjögren's syndrome is usually below 0.5 to 1.0 ml/minute (normal 1 to 1.5 ml/minute).
- *Sialochemistry*—parotid saliva in Sjögren's syndrome contains twice as much total lipid and has elevated content of phospholipids and glycolipids than the normal saliva. The sodium chloride and phospholipids levels are higher in saliva of Sjögren's syndrome patient.
- *Immunologic*—a routine autoantibody profile can usually be carried out with the particular aim of detecting rheumatoid and antinuclear factors.
- *Hematological investigations*—it is necessary, particularly to exclude anemia. ESR or plasma viscosity, leucopenia occasionally may also be found.
- *Microbiological investigations*—a swab from oral mucosa should be taken to confirm candidiasis, if there is soreness and erythema. Examination of pus is also

of course essential as a guide to antimicrobial treatment, if acute sialadenitis develops.

- *Salivary gland biopsy*—the changes in minor glands of lower lip show close correlation with those in the major salivary glands and provide a safe and convenient source of material.

Management

- *Ocular lubricant*—keratoconjunctivitis is treated by instillation of ocular lubricants, such as artificial tears coating methylcellulose and xerostomia is treated by saliva substitutes.
- *Oral hygiene maintenance*—scrupulous oral hygiene and frequent fluoride application is indicated to reduce these problems.
- *Salivary stimulant*—bromhexine, pilocarpine and cevimeline can be used to stimulate salivary flow.
- *Surgery*—surgery for enlargement of salivary gland is only recommended when the enlargement is causing discomfort to the patient.

Mikulicz's Disease or Benign Lympho-epithelial Lesion

It was first described by Mikulicz in 1888 as symmetric or bilateral, chronic, painless enlargement of lacrimal and salivary glands. It exhibits both inflammatory and neoplastic characteristics. Initially, Mikulicz's disease was confused with disease processes such as leukemia and tuberculosis and in these cases, it is called as *Mikulicz syndrome*. Mikulicz's disease is term used only for benign lymphoepithelial lesion involving parotid and lacrimal gland.

Clinical Features

- *Age and sex*—it occurs more commonly in women in middle and later life.
- *Site*—it is manifested as a unilateral or bilateral enlargement of parotid and/or submandibular gland.
- *Prodromal symptoms*—the onset of the lesion is some times associated with fever, upper respiratory tract infection, oral infection, tooth extraction or some local inflammatory disorders.
- *Symptoms*—in some cases, there is mild local discomfort, occasional pain and xerostomia.
- *Signs*—there is often diffuse, poorly outlined enlargement of salivary gland rather than formation of a discrete tumor nodule. The enlargement varies in size but generally few centimeters in diameter. There is history of alternating increases and decreases in the size of mass, from time to time.
- *Duration*—the duration of the tumor mass may be only a few months or many years.

Diagnosis

- *Clinical diagnosis*—unilateral or bilateral enlargement of parotid and lacrimal gland may aid to diagnosis.
- *Laboratory diagnosis*—biopsy shows solid nest or clumps of poorly defined epithelial which termed as 'epimyoepithelial islands'.

Management

- *Surgical excision*—surgical removal of involved gland should be carried out. Prognosis is good.

Uveoparotid Fever

It is a form of sarcoidosis and it is also called as 'Heerfordt's syndrome'.

It consists of a triad of—

- *Uveitis*—inflammation of uveal tract of the eye.
- *Parotid swelling*—firm, painless and bilateral enlargement of parotid gland.
- *Facial palsy*

Etiology

- *Infection*—tuberculosis was earlier thought to be the causative agent.
- *Hereditary*—hereditary factors also play an important role in this syndrome.
- *Autoimmune*—autoimmune mechanism of the body can also be responsible.

Clinical Features

- *Age*—it usually occurs in 3rd and 4th decades of life.
- *Prodromal symptoms*—prodromal symptoms lasting from a few days to several weeks are the usual initial signs of the disease and patient complains of fever, malaise, weakness, nausea and night sweat.
- *Appearance*—it appears as a bilateral, firm, painless parotid swelling. The parotid swelling lasts from several months to several years.
- *Involvement of other gland*—submandibular, sublingual and lacrimal gland swelling may develop independently or during the course of the parotid swelling.
- *Sarcoidial lesion*—sarcoidial lesion may also be found in the oral cavity.
- *Uveitis*—it is an inflammation of the uveal tract, is a feature of this disease. Although ocular symptoms are usually bilateral, they become more apparent before the appearances of parotid swelling.
- *Nerve involvement*—the most common nerve involved is facial nerve. there is unilateral or bilateral seventh nerve paralysis. The neurological signs may precede, follow or appear simultaneously with parotid swelling. There

is trigeminal paresthesia, eyelid ptosis, polyneuritis, intercostal neuralgia and spinal nerve impairment accompanied by weakness and muscle atrophy have been reported.

Radiographic Features

- The sialographic picture merely shows the severity and duration of the disease process within the particular gland.

Diagnosis

- *Clinical diagnosis*—parotid swelling with facial palsy and uveitis will diagnose this syndrome.
- *Laboratory diagnosis*—it will reveal a characteristic sarcoid nodule.

Management

- *Corticosteroids*—it is largely asymptomatic as it may undergo spontaneous remission. Corticosteroid can be used in cases of acute exacerbation.

Recurrent Non-specific Parotitis

It occurs in children as well as in adults, with the average age of onset being between 3 and 4 years. The disease may disappear with puberty, but often progresses into adult life. Males are more commonly affected than females. A congenital defect and allergy have been suggested as the possible causes.

The disease is characterized by a sudden onset of parotid swelling. Both unilateral and bilateral parotid involvement has been reported. Swelling develops rapidly and may persist for a few days to a year. Diminished salivary flow has also been noted during the period of both exacerbation and remission. Moderate rise in total serum protein. Serum gammaglobulin levels may also be increased.

Tumors of Salivary Glands

It is important to note that neoplasms arise not only in major salivary glands, but also in minor salivary glands.

Classification

See Tables 26-1 and 26-2.

Firmness of the Tumor

Firmness results from dense aggregates of nests and cords of closely packed tumor. Fibrous tissue and hyaline area as well as cartilage like and bone like tissue. Softness results from fluid production and its retention phenomenon.

Table 26-1: WHO 1991

Adenoma

- Pleomorphic adenoma
- Myoepithelioma
- Basal cell adenoma
- Warthin's tumor
- Oncocytoma
- Canalicular adenoma
- Sebaceous adenoma
- Ductal papilloma
 - Inverted ductal papilloma
 - Intraductal papilloma
 - Sialadenoma papilliferum
- Cystadenoma
 - Papillary cystadenoma
 - Mucinous cystadenoma
- Monomorphic adenoma
 - Adenolymphoma (Warthin's tumor)
 - Oxyphilic adenoma (oncocytoma)
 - Other types
 - Basal cell adenoma
 - Canalicular adenoma
- Mucoepidermoid tumor
- Acinic cell tumor

Carcinoma

- Acinic cell carcinoma
- Mucoepidermoid carcinoma
- Adenoid cystic carcinoma
- Polymorphous low grade adenocarcinoma
- Epithelial myoepithelial carcinoma
- Basal cell adenocarcinoma
- Sebaceous carcinoma
- Papillary cystadenocarcinoma
- Mucinous adenocarcinoma
- Oncocytic carcinoma
- Salivary duct carcinoma
- Adenocarcinoma
- Malignant myoepithelioma
- Carcinoma in pleomorphic adenoma (Malignant pleomorphic adenoma)
- Squamous cell carcinoma
- Small cell carcinoma
- Undifferentiated carcinoma
- Other carcinomas

Non-epithelial tumors**Malignant lymphoma****Secondary tumors****Unclassified tumors****Tumor like lesions**

- Sialadenosis
- Oncocytosis
- Necrotizing sialometaplasia
- Benign lymphoepithelial lesions
- Salivary gland cyst
- Chronic sclerosing sialadenitis of submandibular gland
- Cystic lymphoid hyperplasia in AIDS

Table 26-2: Clinical

Benign seldom recurrent

- Warthin's tumor
- Oncocytoma
- Monomorphic salivary adenomas

Benign, often recurrent

- Pleomorphic adenoma
- Mucoepidermoid tumor (low grade)
- Acinic cell tumor

Malignant

- Carcinoma in pleomorphic adenoma
- Adenoid cystic carcinoma
- Acinic cell tumor
- Mucoepidermoid tumor (high grade)
- Squamous carcinoma
- Adenocarcinoma
- Undifferentiated carcinoma

Firm tumor	Soft tumor
• Pleomorphic adenoma • Adenoid cystic carcinoma • Mucoepidermoid tumor of high grade • Carcinoma in pleomorphic adenoma • Acinic cell carcinoma • Oncocytoma	• Well differentiated Mucoepidermoid tumor • Papillary cyst adenoma • Mucus producing adenocarcinoma • Warthin's tumor

Clinical Staging of Salivary Gland Tumors**By Spiro**

Staging of salivary gland neoplasms appear to be initiated by Spiro. It is as follows:

- T_1 —0 to 3 cm and solitary and freely mobile and CRVII intact.
- T_2 —3.1 to 6 cm and solitary and freely mobile or skin fixation and CRVII intact.
- T_3 —6 cm or multiple nodules or ulceration or deep fixation or CRVII dysfunction.
- Patient with T_1 and T_2 lesion are placed into stage I and II respectively.
- Any patient with clinical evidence of metastasis of lymph nodes or with T_3 lesion is considered to be in stage III.

By American Joint Committee**Primary tumor**

- T_x —tumor that cannot be assessed by the rules.
- T_0 —no evidence of primary tumor.
- T_1 —tumor 2 cm or less in diameter, without significant local extension.
- T_2 —tumor 2-4 cm in diameter without significant local extension.
- T_3 —tumor more than 4 cm but not more than 6 cm in diameter without significant local extension.

- T_{4a} —tumor over 6 cm in diameter without significant local extension.
- T_{4b} —tumor of any size with significant local extension.
- *Nodal involvement (N)*
 - N_x —regional lymph node cannot be assessed.
 - N_0 —no regional lymph node metastasis
 - N_1 —clinical or histologically positive regional lymph nodes.
- *Distant metastasis (M)*
 - M_x —distant metastasis cannot be assessed.
 - M_0 —no distant metastasis.
 - M_1 —distant metastasis.
- *Stage grouping is performed as follows.*
 - *Stage I*— $T_1N_0M_0$ or $T_2N_0M_0$
 - *Stage II*— $T_3N_0M_0$
 - *Stage III*— T_1 or $T_2, N_1 M_0$, or T_{4a} or $T_{4b} N_0 M_0$
 - *Stage IV*— $T_3 N_1 M_0$, T_{4a} or $T_{4b} N_1 M_0$, any T any N M_1

Benign Tumors

Pleomorphic Adenoma

The term pleomorphic adenoma was suggested by Willis characterizing the unusual histological pattern of the lesion (pleomorphic or mixed appearance). It is a benign mixed tumor of the salivary gland. It is also called as 'iceberg tumor', 'endothelioma', 'branchioma', or 'enchondroma'. It is most common salivary gland tumor.

Pathogenesis

- *Myoepithelial cells*—myoepithelial cells are responsible for morphologic diversity of tumor including production of fibrous, mucinous, chondroid and osseous structures.
- *Differentiation of ductal reserve cells*—intercalated duct reserve cells can differentiate into ductal and myoepithelial cells and the later can then undergo mesenchymal metaplasia since they inherently have smooth muscle like properties.
- *Neoplastic altered epithelial cells*—a neoplastically altered epithelial cell with potential for multidirectional differentiation may be responsible for pleomorphic adenoma.

Clinical Features

- *Sex and age*—women to men ratio is 6:4. It is common in 4th to 6th decades but also seen in young adults and children
- *Site*—parotid 90% and intraoral palatal gland on lip. In parotid involvement, superficial portion is most commonly affected (Fig. 26-23).



Fig. 26-23: Swelling of parotid gland seen in cases of pleomorphic adenoma.

- *Symptoms*—small, painless, quiescent nodule which slowly begins to increase in size, sometimes intermittently.
- *Shape*—the tumor tends to be round or oval when it is small, as it grows bigger it becomes lobulated. In some cases, intraorally swelling may be observed (Fig. 26-24).



Fig. 26-24: Intraoral view pleomorphic adenoma showing swelling in the palate (Courtesy Dr Parate).

- *Size*—it may increase to cricket ball size or even more, weighing in pounds and in intraoral cases, not more than 1 to 2 cm in diameter. If neglected lesion can grow bizarre proportions.
- *Dumbbells shaped appearance*—in some cases, lesion grows in medial direction between ascending ramus and stylomandibular ligament resulting in dumbbell shaped appearance.
- *Surface*—its surface is smooth. Sometime, it is bosselated and is occasionally crossed by deep furrows.

- *Fixation*—no fixation, either to deeper tissues or overlying skin.
- *Consistency*—it is firm and rubbery to feel. Sometimes cystic degeneration may be seen.
- *Palatal tumor*—it is seen on lateral aspect of the palate. They are smooth surface and dome shaped (**Fig. 26-25**).
- *Signs of malignant transformation*—accelerated growth rate, tumor irregularity on palpations, necrosis and painful ulceration and facial nerve involvement.



Fig. 26-25: Dome-shaped swelling on lateral aspect of palate due to pleomorphic adenoma of the minor salivary gland (Courtesy Dr Chole).

Diagnosis

- *Clinical diagnosis*—smooth surface enlargement in the parotid region will suspect pleomorphic adenoma.
- *Sialography*—ball in hand appearance seen (**Fig. 26-26**).
- *CT diagnosis*—this will also help to know exact extension of location (**Fig. 26-27**).
- *Laboratory diagnosis*—biopsy shows cuboidal cells arranged in tubes or duct-like structures, which begin to resemble the normal ductal epithelium. Duct-like spaces contains eosinophilic coagulum.

Management

- *Surgical excision.*
 - *Parotid gland*—tumor and the involved lobe of gland is removed.
 - *Submaxillary gland*—removal of the gland and tumor in continuity.
 - *Intraoral lesion*—extracapsular incision.
 - *Hard palate*—excised with overlying mucosa.
 - *Lip, soft palate*—enucleation or extracapsular excision.
- *Recurrence rate*—recurrence rate is 5 to 30% due to hypocellularity, incomplete resection and encapsulation.

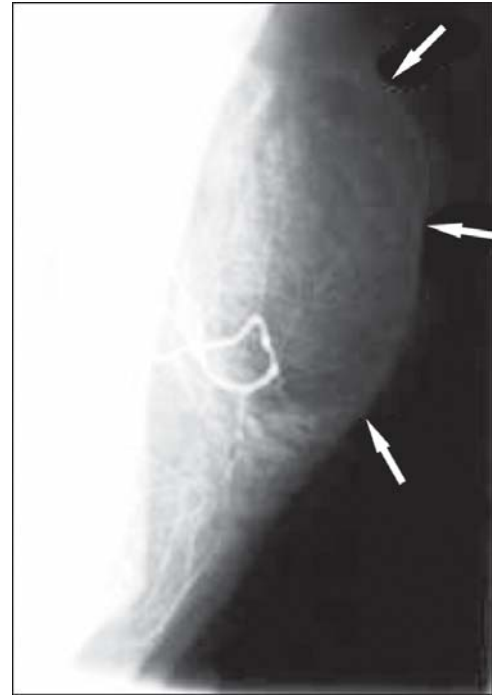


Fig. 26-26: Ball in hand appearance seen on sialography in pleomorphic adenoma (Courtesy M Shimizu).

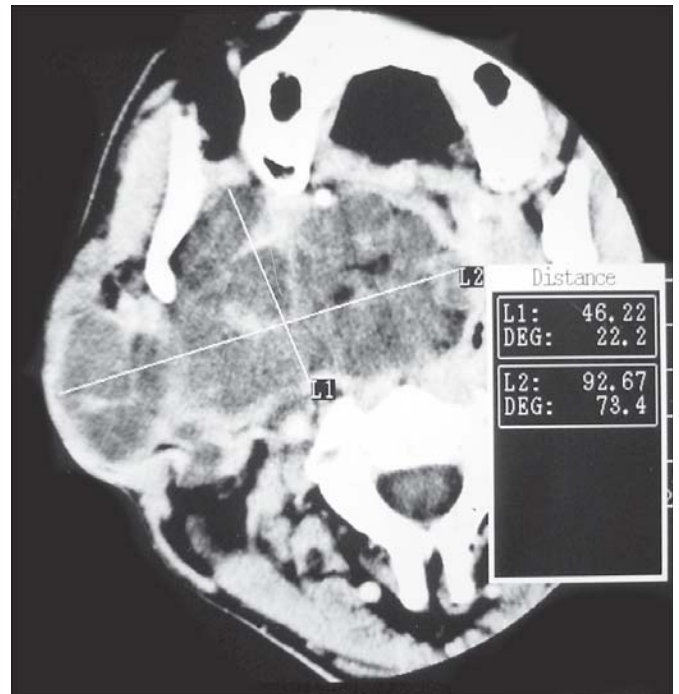


Fig. 26-27: CT scan showing extension of pleomorphic adenoma involving parotid gland

Monomorphic Adenoma

Monomorphic adenoma is divided into three groups by WHO, i.e. adenolymphoma (Warthin's tumor), Oxyphilic adenoma (oncocyoma) and other histologic patterns like basal cell adenoma and canalicular adenoma. But

Warthin's tumor and Oxyphilic adenoma are recognized separate entity nowadays; so only included in monomorphic adenoma are basal cell adenoma and canalicular adenoma.

Basal Cell Adenoma

It derived its name by baseloid appearance of the tumor cells.

Clinical Features

- *Age and sex*—it is more common in females. Older age group, usually over 60 years of age are affected.
- *Site*—it occurs primarily in major salivary glands particularly in the parotid gland and intraorally, upper lip.
- *Symptoms*—the tumor is usually painless and is characterized by slow growth.
- *Membranous basal cell adenoma*—this type of variant of basal cell adenoma occurs in association with appendage tumors like dermal cylindroma and trichoepitheliomas.

Diagnosis

- *Clinical diagnosis*—it is difficult to make clinical diagnosis.
- *Laboratory diagnosis*—biopsy shows fairly well defined connective tissue capsule. The cells are isomorphic and baseloid with round to oval nuclei.

Management

- *Surgical excision*—it is treated by surgical excision and recurrence is seldom seen.

Canalicular Adenoma

It occurs exclusively in minor salivary gland.

Clinical Features

- *Age and sex*—it is common in patient over the age of 60 years with no sex predilection.
- *Site*—it originates primarily in the intraoral accessory glands. It occurs in upper lips followed by palate, buccal mucosa and lower lip.
- *Symptoms*—it presents as a slowly growing, well circumscribed, firm nodule.
- *Signs*—it is firm and fluctuant to palpation.

Diagnosis

- *Clinical diagnosis*—firm fluctuant swelling on upper lip may suspect the diagnosis of canalicular adenoma.

- *Laboratory diagnosis*—biopsy shows long strands or cords of epithelial cells, arranged in a double row. It shows 'party wall'.

Management

- *Surgical excision*—it can be treated by simple enucleation and surgical excision. Recurrence rate is rare.

Warthin's Tumor

It is also called as 'adenolymphoma' and 'primary cystadenoma lymphomatosum'. This tumor was first described by Albrecht and Arzt but it usually bears the name of Warthin in the recognition of the pathologist who first described it in USA in 1929.

Development

- *Heterotrophic salivary gland tissue*—tumor arises from salivary gland tissue entrapped with para-parotid or intra-parotid lymph nodes during embryogenesis.
- *Delayed hypersensitivity disease*—it is most likely a delayed hypersensitivity disease, the lymphocytes being an immune reaction to the salivary ducts which undergo oncocytic change.
- *Secretory immune response*—Hsu has suggested that it is an exaggerated secretory immune response.
- *Other factors*—other factors like smoking and Epstein-Barr virus have also been responsible for Warthin tumor.

Clinical Features

- *Age and sex*—it is common in men (male to female ratio is 5:1). It is common in 6th decade.
- *Site*—the tumor occurs almost exclusively in the parotid gland. It always occurs in the lower portion of the parotid gland. The tumor is generally superficial, lying just beneath the parotid capsule or protruding through it.
- *Symptoms*—the usual complaint is painless slow growing tumor over the angle of jaw. Involvement may be bilateral or may be multifocal.
- *Size and shape*—the tumor does not attain a large size and the usual size is 1-3 cm in diameter. It is spherical in shape.
- *Surface*—it is smooth (**Fig. 26-28**) and it is well circumscribed, movable.
- *Consistency*—it classically feels doughy and compressible on palpation. It is firm on palpation and is clinically indistinguishable from other benign lesions of parotid gland.

Radiological Features

- *CT/MRI features*—Warthin tumor is observed as a round or oval shaped mass with smooth, well-delineated

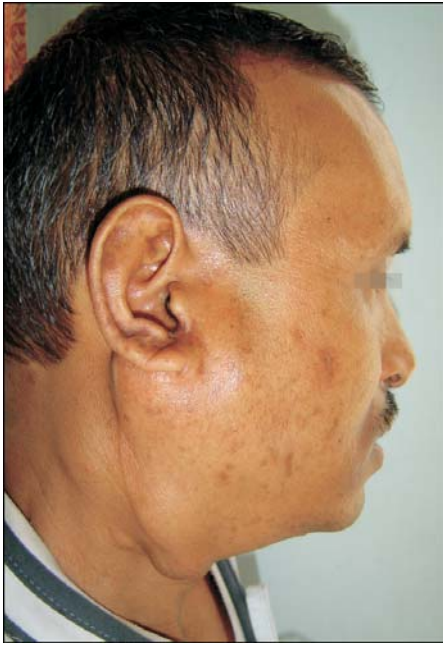


Fig. 26-28: Smooth swelling occurs due to mass in tail of parotid gland in Warthin tumor (Courtesy Dr Chole).

contour. It contains cystic parts, which are seen on CT, MR images (**Fig. 26-29**) and sonograms.

- **Sonography**—on sonograms, this tumor is observed as a very clearly delineated mass with multiple cystic areas. On Doppler mode, relatively abundant vascularization is observed.



Fig. 26-29: Warthin tumor: T1 weighted MR image shows round shaped mass with smooth, well-delineated contour in the lower pole of the left parotid gland. Signal intensity is higher than that of muscle (Courtesy Dr Shimizu).

Diagnosis

- **Clinical diagnosis**—difficult to diagnose.
- **Laboratory diagnosis**—it is an adenoma exhibiting cyst formation, with papillary projections into the cystic spaces and lymphoid matrix showing germinal centers.

Management

- **Superficial parotidectomy**—it is done as tumor is usually located in superficial part of the parotid gland.

Oncocytoma

It is also called as '*oxyphilic adenoma*', '*acidophilic adenoma*'. It is an uncommon tumor composing less than 1% of salivary neoplasms. The term oncocytoma is derived from the resemblance of these tumor cells to apparently normal cells, which have been termed as '*oncocytes*'. These cells are predominately seen in duct lining of glands in elderly persons.

Clinical Features

- **Age and sex**—it is more common in women than in men and occurs almost exclusively in older persons.
- **Site**—it usually occurs in the parotid gland.
- **Size**—the tumor usually measures 3 to 5 cm in diameter and appears as a discrete encapsulated painless mass (**Fig. 26-30**) which is sometimes nodular. Pain is generally absent.
- **Oncocytosis**—an interesting condition called '*oncocytosis*' of parotid gland has been described. It is characterized by nodules of oncocytes involving the entire gland or a large portion. It is usually bilateral.



Fig. 26-30: Discrete encapsulated painless mass seen in oncocytoma (Courtesy Dr Parate).

Diagnosis

- *Clinical diagnosis*—it is difficult to differentiate with other benign condition.
- *Laboratory diagnosis*—biopsy shows large cells with eosinophilic cytoplasm, distinct cell membrane. It is arranged in rows and cords.

Management

- *Surgical excision*—surgical excision of tumor should be carried out. Facial nerve should be preserved.

Myoepithelioma

It is an uncommon salivary gland tumor. It occurs in adults and has equal sex distribution. Parotid gland is most commonly involved and the palate is the most frequent intraoral site of occurrence. The clinical features are same as pleomorphic adenoma.

The tumor is composed of spindle shaped or plasmacytoid cells or combination of the two cell types. These cells may be set in myxomatous background, which may vary from scanty to copious. It is managed by surgical excision.

Ductus Papillomas

These are the group of tumor characterized microscopically by papillomatous pattern.

Types

- *Simple or intraductal papilloma*—appears as submucosal swelling.
- *Inverted ductal papilloma*—is present most commonly on lip.
- *Sialadenoma papilliferum*—is seen most commonly on palate (**Fig. 26-31**).



Fig. 26-31: Ductus papilloma presenting as swelling on hard palate.

Clinical Features

- *Simple or intraductal papilloma*—it is present as an exophytic lesion with a papillary surface and is pedunculated. It is usually reddish in color and present on the buccal mucosa or palate. It is ill-defined.
- *Inverted ductal papilloma*—it is present asymptomatic nodules of the oral mucosa of adults. It is more commonly seen on lip. It may show pit or indentation in the overlying surface mucosa.
- *Sialadenoma papilliferum*—the lesion occurs in adults as an exophytic papillary lesion of the hard palate.

Diagnosis

- *Clinical diagnosis*—it is difficult to make. But small swelling in relation with minor salivary gland, one should suspect ductal papilloma.
- *Laboratory diagnosis*—on biopsy, it shows multiple exophytic papillary projection which is covered by stratified squamous epithelium.

Management

- *Surgical excision*—it is best treated by conservative surgical excision.

Malignant Tumors

Peripheral Mucoepidermoid Carcinoma

The term mucoepidermoid tumor was introduced in 1945 by Stewart, Foote and Becker. It accounts for 6 to 9 % of the salivary gland tumors and for about 1/3rd of all malignant tumors of the salivary glands. It consists, of both, mucus secreting as well as epidermoid type of cells as its name suggests.

It is of mainly two types, i.e. benign and malignant based upon clinical nature and histologic feature of the lesion.

Clinical Features

- *Age and sex*—it occurs in equal distribution in men and women and occurs in the 3rd and 5th decade.
- *Site*—about 60% occur in parotid gland (**Fig. 26-30**) and 30% in the minor salivary glands, especially those of the palate. It can be seen on buccal mucosa, tongue and retromolar area.
- *Onset*—it appears as a slowly enlarging painless mass, which simulates pleomorphic adenoma (**Figs 26-32A and B**).
- *Symptoms*—pain and facial nerve palsy can be seen in some patients. Other findings such as lacrimation, trismus, nasal discharge, and blood tinged saliva can also be seen.



Figs 26-32A and B: Mucoepidermoid carcinoma of parotid gland. (A) Extraoral view, (B) Intraoral view (Courtesy Dr Parate).

- *Low grade tumor*—the tumor of low grade malignancy usually appears as a slowly enlarging, painless mass, which stimulates pleomorphic adenoma. It seldom exceeds 5 cm in diameter (**Fig. 26-33**).
- *High grade malignancy*—the tumor of high grade malignancy grows rapidly and does produce pain as an early symptom. It tends to infiltrate the surrounding tissues and in high percentage of cases it metastasizes to the regional lymph nodes. Distant metastases to lungs, bones, brain and subcutaneous tissue are common. In some cases, extraoral ulceration can also be seen (**Fig. 26-34**).
- *Minor gland tumors*—it appears as asymptomatic tumors which is blue or red in color.

Diagnosis

- *Clinical diagnosis*—it is difficult to make clinical diagnosis of mucoepidermoid carcinoma.

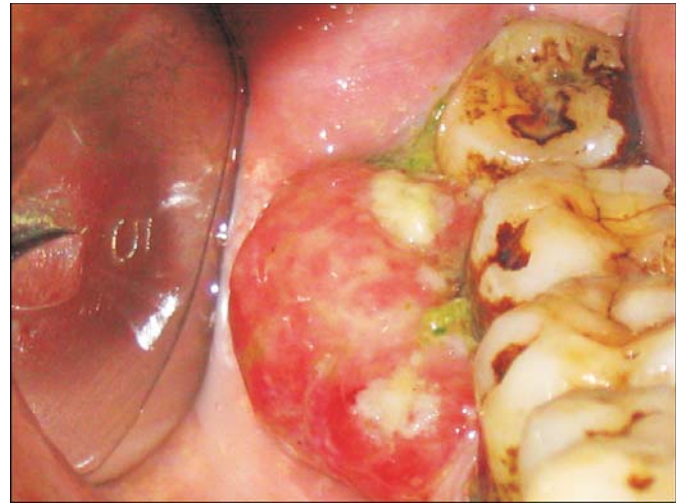


Fig. 26-33: Mucoepidermoid tumor occurring on buccal mucosa (Courtesy Dr Soni).



Fig. 26-34: Mucoepidermoid carcinoma showing extraoral ulceration

- *Radiological diagnosis*—computed tomography will diagnose this condition (**Fig. 26-35**).
- *Laboratory diagnosis*—biopsy shows mucus-secreting cells, epidermoid type cells and intermediate cells. Biopsy also shows sheets or nests of epidermoid cells and similar nests of mucus cells, usually arranged in a glandular pattern and showing microcyst formation.

Management

- *Subtotal parotidectomy*—early stage tumor is managed by subtotal parotidectomy and facial nerve preservation.
- *Total removal*—advanced cases need total removal of the parotid gland.

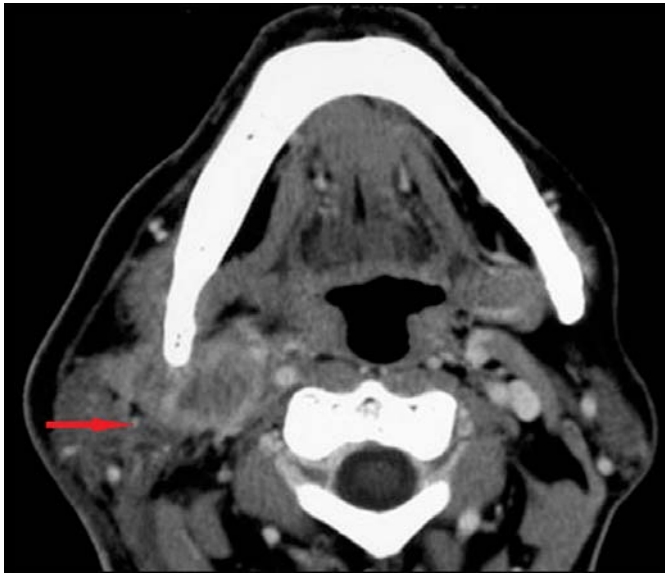


Fig. 26-35: Computed tomography showing growth on right side in parotid gland (arrow).

- *Radical neck dissection*—it is indicated for the person who has clinical evidence of metastasis.
- *Radiotherapy*—it can be given in some cases of intermediate type of mucoepidermoid carcinoma.

Central or Intraosseous Mucoepidermoid Carcinoma

It is an epithelial tumors originating in the bone and is believed to be derived from salivary gland.

Origin

- *Entrapment of retromolar mucus gland*—it may originate from entrapment of retromolar mucus glands within the mandible, which subsequently undergo neoplastic transformation.
- *Embryonic remnants of submaxillary gland*—it may also form from developmentally retained embryonic remnants of the submaxillary gland within the mandible and neoplastic transformation of the mucus secreting cells commonly found in the epithelial lining of dentigerous cyst.

Clinical Features

- *Age and sex distribution*—it is most commonly seen in middle age individual. It is more common in females than males.
- *Site*—it occurs in mandible in premolar-molar area. It does not extend anteriorly beyond the premolar region.
- *Symptoms*—patient complains of painless swelling. There may be paresthesia of inferior alveolar nerve.
- *Sign*—swelling may cause facial asymmetry. Tenderness is present. Regional lymph nodes are enlarged.

Radiological Features

- *Site*—it occurs in premolar-molar area of mandible, above mandibular canal.
- *Appearance*—it presents as a unilocular or multilocular expansile mass (Fig. 26-36).
- *Margin*—it is most often well defined, well corticated and often crenated or undulating in nature.
- *Soap bubble or honeycomb appearance*—lesion may have soap bubble or honeycomb internal structure.



Fig. 26-36: Multilocular lesion seen in premolar molar area (Courtesy Dr Parate).

- *Effect on surrounding structure*—the buccal and lingual cortical plate, inferior border of the mandible, alveolar crest may be thinned and grossly displaced. The mandibular canal is depressed or pushed laterally or medially. Lamina dura of the teeth may be lost.

Diagnosis

- *Clinical diagnosis*—not so specific.
- *Radiological diagnosis*—soap bubble and honeycomb appearance may be seen.

Differential Diagnosis

- *Ameloblastoma*—radiologically, it is difficult to differentiate. Diagnosis should be confirmed by histopathological investigation.
- *Odontogenic myxoma*—there is history of missing tooth.
- *Central giant cell granuloma*—it frequently crosses the midline.

Management

- *Surgical excision*—it is treated by surgical excision of tumor.
- *Neck dissection*—neck dissection should be carried out in case of metastatic.

- *Adjunctive radiotherapy*—postoperative radiation therapy may be required for control of metastasizing disease.

Acinic Cell Tumor

It is also called as '*acinic cell or serous cell adenoma*'. It accounts for approximately 1% of all salivary gland tumors. It shows serous acinar differentiation.

Clinical Features

- *Age and sex*—it occurs in middle age and twice common in women.
- *Site*—it arises exclusively in the superficial lobe and tail of the parotid gland. The most common intraoral sites are the buccal mucosa and lip.
- *Symptoms*—it is painless and grows slowly.
- *Signs*—exact delineation of the lesion is difficult and attachment to the overlying skin and muscle may occur.
- *Progress*—some of these lesions run a rapid course with hematogenous and lymphatic metastases, while others are more slowly progressive. Locally invasive growth may be encountered in some lesions.

Diagnosis

- *Clinical diagnosis*—not possible.
- *Laboratory diagnosis*—biopsy shows thin capsule, which composed of cells of varying degrees of differentiation. Well differentiated cells bear remarkable resemblance to normal acinar cells, whereas less differentiated cells resemble embryonic ducts and immature acinar cells.

Management

- *Lobectomy*—superficial lobe of the parotid gland should be excised, is the treatment of choice. Recurrence rate varies from 8 to 59%.

Adenoid Cystic Carcinoma

It is also called as '*cylindroma*', '*adenocystic carcinoma*' and '*baseloid mixed tumor*'. It is best recognized salivary gland malignancy.

Clinical Features

- *Age*—it occurs in the 5th and 6th decade of life.
- *Site*—most common glands involved are the parotid, sub-maxillary and the accessory glands in palate and tongue.
- *Symptoms*—the most common initial symptom is presence of mass followed by local pain, facial nerve paralysis in case of parotid tumor and tenderness.
- *Signs*—some of the lesions exhibit surface ulceration. Other findings include nasal obstruction, proptosis,

sinusitis, ear infection, epistaxis, signs of cranial nerve involvement and visual disturbances.

- *Metastasis*—the incidence of metastases is more and the organs involved include cervical lymph nodes, lungs, brain, liver and kidneys.

Radiological Features

- *Appearance*—CT scan will demonstrate destructive lesion (Fig. 26-37).

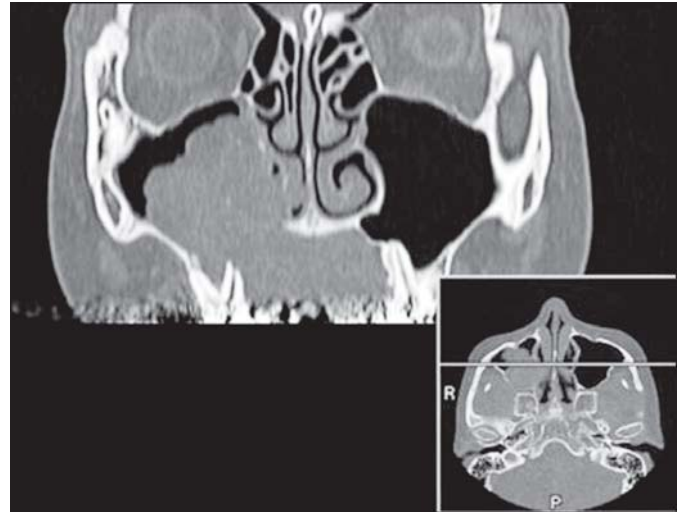


Fig. 26-37: This tumor shows bone destruction and absorption of the dental root. Some fine calcification can also be seen in the tumor (Courtesy Dr Shimizu).

Diagnosis

- *Clinical diagnosis*—not possible.
- *Laboratory diagnosis*—biopsy shows basal cells arranged in anastomosing cords or may contain a mucoid material producing the typical cribriform '*honeycomb*' or '*Swiss cheese*' pattern. The stromal connective tissue becomes hyalinized and surrounds the tumor cells, forming a structural pattern of cylinder from which the lesion originally derived the name '*cylindroma*'.

Management

- *Surgical excision*—surgical excision is the treatment of choice. Recurrence rate is about 60 to 92%. Long-term follow up is hence essential.
- *Adjunct radiotherapy*—surgery should be accompanied by radiation therapy.

Adenocarcinoma

It is a malignant epithelial tumor showing tubules or papillary glandular formation. All adenocarcinoma are

highly malignant and metastasize to regional lymph nodes and general viscera. Computed tomography will diagnose this malignant tumor (**Fig. 26-38**).

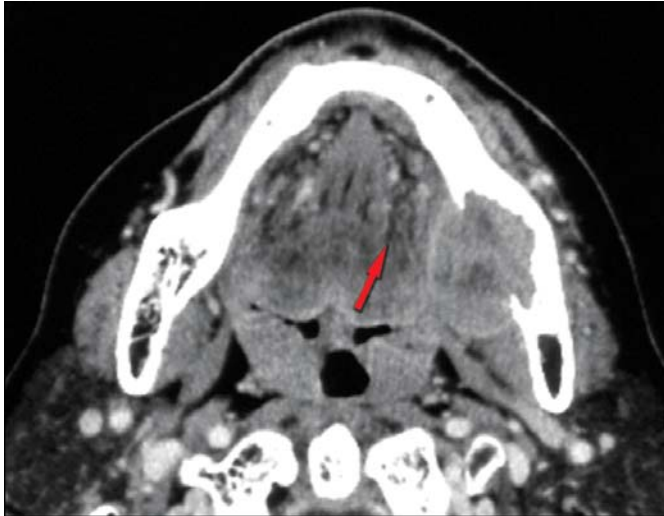


Fig. 26-38: Enhanced CT shows round, slightly enhanced mass with bone invasion in the lingual side of the left retro-molar region. Bone invasion suggests malignant nature of this tumor

Malignant Pleomorphic Adenoma

It is also called as '*malignant mixed tumor*'. It is uncertain that these tumors represent the previously benign lesions which have undergone transformation into malignant form or are malignant lesion right from the onset. It accounts for 1% of all parotid tumors and 7% of all malignant tumors.

Clinical Features

- **Age**—the tumor occurs from 2nd to 9th decades, but most frequently in 5th and 6th decade. The average age of patients with malignant pleomorphic adenoma is about ten years older than the patients with benign form of the disease.
- **Symptoms**—pain is more frequently a feature of malignant, than the benign pleomorphic adenoma
- **Size**—the tumors are usually larger than benign ones.
- **Signs**—there is often fixation of the tumor to underlying structures as well as to overlying skin or mucosa.

Radiological Features

- **CT features**—CT/MRI features will show malignant changes in the tumors (**Fig. 26-39**).

Diagnosis

- **Clinical diagnosis**—not possible.
- **Laboratory diagnosis**—biopsy shows malignant component. There is presence of nuclear hyperchromatism and pleomorphism, increased or abnormal mitosis and destruction of normal tissues.

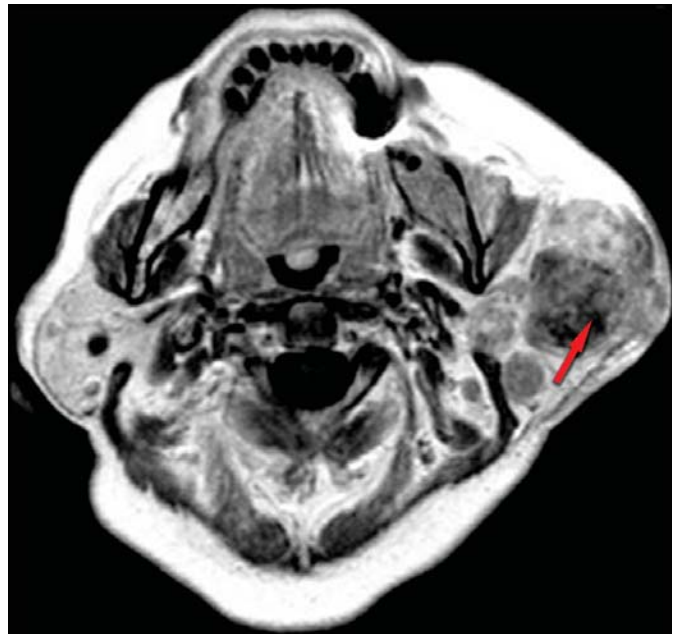


Fig. 26-39: T1 weighted MR image shows several masses in the left parotid gland. The relatively well-delineated mass in the middle shows low signal intensity (Courtesy Dr Shimizu).

Management

- **Surgical**—surgical excision with local node dissection should be carried out. These neoplasms exhibit a high recurrence rate after surgical removal as well as a high incidence of regional lymph node involvement.

Epidermoid Carcinoma

It is also called as '*squamous cell carcinoma of salivary glands*'. It is thought to be ductal in origin, since duct may undergo squamous metaplasia with ease. It exhibits infiltrative properties and early metastasis. It recurs readily. It is not a common lesion and arises more frequently in the major salivary gland. The combined use of surgery and radiotherapy is more beneficial.

Undifferentiated Carcinoma

The tumor is composed of round or spindle cells that are poorly differentiated. Undifferentiated carcinomas infiltrate the surrounding structures and readily metastasize to cervical lymph nodes.

Metastatic Carcinoma

Salivary glands may be a site for tumor metastases, since the parotid gland contains as many as 20 to 30 lymph follicles. Parotid and paraparotid lymph nodes are favored sites for metastases from malignancy of the temple, scalp and ear and occasionally from the face, neck and palate. The most common metastasizing tumors are melanoma and squamous cell carcinoma.

Connective Tissue Tumors

Hemangioma is the only common tumor of this group and the most frequently seen in young infants. The involved glands appear hypertrophied. There is blue discoloration of the overlying skin. Lipoma may occur in the parotid gland. The facial nerve, occasionally gives rise to a neural tumor. True sarcoma is extremely rare.

Necrotizing Sialometaplasia

It is a non-neoplastic inflammatory self-healing reaction of salivary gland tissues, which both clinically and histologically mimics a salivary gland malignancy. It usually affects the minor salivary glands. It was predicted that trauma causes ischemia of the minor salivary glands. This benign self-limiting lesion has been often confused with malignancy thereby causing unnecessary surgery.

Etiology and Pathogenesis

- *Local ischemia*—it may be due to physical, chemical, infective or local vasculitis.
- *Trauma*—trauma from various factors such as denture wearing and recent surgery may be a causative factor.
- *Other factors*—in some cases, smoking and alcohol intake can lead to this condition. It can also occur in upper respiratory tract infection.

Clinical Features

- *Age and sex*—it is more common in males than females. Occurs in 4th and 5th decade.
- *Site*—most of the cases occur in palate, although cases of lip or retromolar pad also have been reported.
- *Onset*—there may be early mild swelling which appears as non-ulcerated swelling (**Fig. 26-40**).



Fig. 26-40: Bilateral swelling seen in palatal region due to necrotizing sialometaplasia. Slight ulceration seen on the swelling.

- *Symptoms*—it is usually painless or may cause only slight pain. The patient may have numbness in the palate, or area of 'looseness' in palate. Pieces of tissue may fall out from the palate. There may be referred pain to ear or pharynx.
- *Signs*—the lesion begins as a larger ulcer or ulcerated nodule, which may be unilateral or bilateral. The ulcer is well demarcated from surrounding normal tissue and often has an inflammatory reaction around the edge of the lesion. Margins of ulcer in some cases, may be indurated and inflamed. The lesion is covered by an inflammatory exudate and necrotic debris, however, the margins of ulcer are usually clean, sharp and the granulation tissue often can be seen at the superficial aspect.
- *Size*—the lesion itself is most often a deep, craterlike, non-draining ulcer 1 to 3 cm in diameter.

Diagnosis

- *Clinical diagnosis*—ulcerative swelling in the palate will aid in the diagnosis.
- *Laboratory diagnosis*—in biopsy, it shows acinar necrosis and squamous metaplasia of salivary ducts.

Differential Diagnosis

- *Traumatic*—short duration and history.
- *Aphthous ulcer*—short duration, painful and heals in 2 to 3 weeks.
- *Squamous cell carcinoma*—rare on palate, older age and biopsy.
- *Syphilis*—painless indurated edema, painless lymph node swelling and serology.
- *Non-Hodgkin's lymphoma*—biopsy.

Management

- *Self limiting*—it is a self-limiting condition. It does not require any treatment. The healing occurs in six to twelve weeks via secondary intention.
- *Debridement*—debridement and saline rinses may aid in the healing process.

Minor Salivary Gland Tumor

Early minor salivary gland tumor is usually nodular or dome, shaped elevation, with smooth contour. Overlying mucosa is normal or appears smooth and glossy, due to tension. After trauma, ulcer appears which is persistent and becomes necrotic. Its occurrence in an posterior aspect of hard palate; next commonly on upper lip, buccal mucosa, retromolar region, tongue and floor of mouth.

There is higher frequency in women with age 30 to 39 for benign and 40 to 49 for malignant tumors.

Mucoepidermoid and adenocarcinoma are common variety. Lung is the common site of distinct metastasis.

Radiologically, it may present well defined saucer like depression of underlying bone in contrast to malignant tumor which invades the bone and produce ragged radiolucent defect.

On sialography, an area of under filling within the gland, due to ductal compression by the tumor is seen. The ducts adjacent to the tumor are usually stretched around this is known as '*ball in hand*' appearance. Retention of contrast media in the displaced ducts during the emptying phase is seen.

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Introduction

Paranasal sinuses are the air filled spaces present with some bone around the nasal cavities. The sinuses are frontal, maxillary, sphenoidal and ethmoidal. Because of the close proximity of the maxillary teeth with the maxillary sinuses, these are the most important paranasal sinuses in dental point of view. They are the largest air filled sinuses surrounding the nose.

Development of Maxillary Sinus

- *Formation of maxillary process*—during 4th week of embryonic life, the dorsal portion of the 1st pharyngeal arch forms maxillary process. It extends forwards beneath the developing eye and gives rise to the maxilla, the zygomatic bone and part of the temporal bone.
- *Evagination from primitive ethmoid infundibulum*—the maxillary sinus starts to develop at about 12 weeks as an evagination from the primitive ethmoid infundibulum in the lateral wall of the middle meatus of the corresponding nasal cavity when the nasal epithelium invades the maxillary mesenchyme.
- *Sinus at birth*—at birth, maxilla is filled with deciduous tooth germs which are very close to the orbital floor so the superior dental nerves and vessels have very short distance to travel to reach the teeth.
- *Separation of alveolar and orbital aspect*—the alveolar and orbital aspects of the maxilla gradually become separated by the cancellous bone which is then resorbed as the sinus enlarges.
- *Pneumatization of maxilla*—pneumatization of the maxilla commences just below the orbital floor.
- *Expansion of maxillary sinus*—down growth of the maxillary sinus leaves the ostium in a position

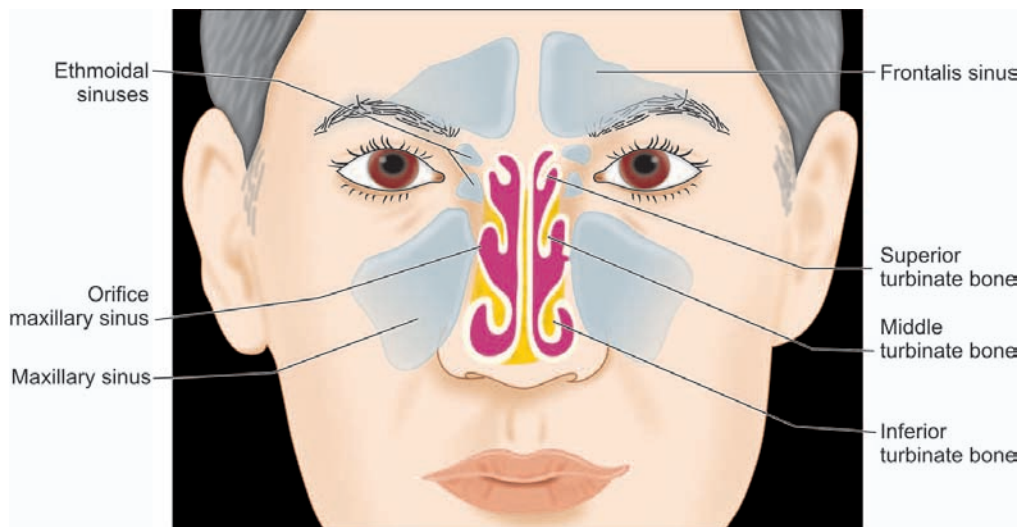


Fig. 27-1: Position of maxillary sinus.

unfavorable for gravitational drainage. The maxillary sinus expands not only downward but also forward and backward from its initial evagination, the site which persists as the antranasal duct. It undergoes concurrent lateral expansion and by the end of the first year, extends beneath the orbit as far as the infraorbital canal (Fig. 27-1).

- *Sinus at the end of 2nd year*—by the end of 2nd year, the sinus has reached about half to its adult size.
- *Facial growth*—as the facial growth is continued by surface deposition on the face, alveolar processes and palate, it is accompanied by the resorption of the internal structure of the maxillary sinus.

Anatomy of Maxillary Sinus

It is also called as the 'antrum of Highmore'. They are the paired structures located largely in the body of maxilla and are mirror image of each other. Maxillary antrum contains air and is lined by mucoperiosteum with a pseudostratified ciliated columnar epithelium.

- *Shape*—it is pyramidal in shape with its base directed medially towards the lateral wall of the nose and the apex directed laterally in the zygomatic process of the maxilla.
- *Size*—size is variable, varying up to 3.5 cm in height and 2.5 cm in width and 3.2 cm anteroposteriorly.
- *Hiatus semilunaris*—it opens into the middle meatus of the nose in the lower part of the *hiatus semilunaris*.
- *Roof*—it is formed by the floor of the orbit and is transversed by the infraorbital nerve. The roof is flat and slopes slightly anteriorly and laterally. The most medial part of the roof forms the sloping wall of the maxilloethmoidal sinuses, from which disease may spread to the maxillary sinus. Antral infection may involve infraorbital vessels and nerve and malignant tumors growing in the sinus may involve the orbit.
- *Floor*—it is curved rather than flat and is formed by the alveolar processes of maxilla and lies about 1 cm below the level of the floor of the nose. Roots of upper molars and premolar may ridge the floor or project into it. Floor may be subdivided by incomplete bony septa lying between the roots of the teeth, especially in the posterior part of the sinus. The 1st and 2nd molar's roots are the most commonly present in close proximity with the maxillary sinus followed by the premolars and 3rd molar.
- *Medial wall*—it is bounded by the nasal cavity. Medial wall is generally slightly convex toward the sinus.
- *Posterior wall*—it is related to the pterygopalatine fossa. The posterior wall bulges posteriorly towards the infratemporal fossa.
- *Lateral wall*—it is related to zygoma and cheek.
- *Anterior wall*—it is related to the cheek. It is depressed by canine fossa on the anterior surface of the maxilla and is convex toward the interior of the sinus.
- *Radiological anatomy*—an antrum appears radiographically as a radiolucent cavity in the maxilla, with well defined, dense, corticated radiopaque margins or walls. The internal bony septa and blood vessel canals in the walls all produce their own shadow.
- *Arterial supply*—facial, infraorbital and greater palatine arteries.
- *Venous drainage*—into the facial and pterygoid plexus of veins.
- *Lymphatic drainage*—drains into the submandibular nodes.
- *Nerve supply*—it is supplied by infraorbital and anterior, middle and posterior superior alveolar nerves.

Function of Maxillary Sinus

- *Reduction of weight of the facial skeleton*—as the maxillary sinus is filled with air rather than cancellous bone, lightens the face by approximately the weight of a pair of spectacles.
- *Phonetic resonance and auditory feedback*—the sinuses may act as a resonating box for the singing voice. The sinuses also affect the conductance of the voice to one's ear.
- *Insulation*—the sinuses may insulate the orbits from intranasal temperature variations.
- *Air conditioning*—the maxillary sinus contain some serous gland whose watery secretion evaporates to humidify the contained air.
- *Water conservation*—the sinuses may act as accessory heat exchanges, warming inspired air to increase its moisture content.
- *Olfaction*—pneumatization may have evolved to increase the area of olfactory mucosa thereby improving the sense of smell.

Examination and Investigation of Maxillary Sinus

- *Clinical examination*
 - *Examination of middle third of face*—the middle third of the face should be inspected for the presence of asymmetry, deformity, swelling, erythema, ecchymosis or hematoma. Epiphora, nasal obstruction, epistaxis or other discharge or odor from the nostril should be noted.
 - *Extraoral palpation*—examination should include palpation of the facial wall of the sinus above the premolar, where the bone is thinnest, either through the soft tissue of the cheek or more directly intraorally.

- *Intraoral examination*—intraoral examination should be performed for alveolar ulceration, expansion, tenderness or paresthesia of the upper molar and premolar region. Palatal ulceration should be biopsied if there is no obvious cause and it does not heal within 1 week.
- *Rhinascopy*—a nasal speculum and headlight or mirrors are necessary for proper examination of the nasal passage. A topical vasoconstrictor may decrease the size of a large inferior turbinate which obstruct the view of the middle turbinate and middle meatus. When the middle meatus is exposed, the hiatus semilunaris and bulla ethmoidalis are seen anteriorly and it is possible to cannulate or probe the ostium more posteriorly. Pus may be seen in middle meatus.
- *Nasendoscopy*—under local anesthesia, the nasal opening of the maxillary sinus in the middle meatus may be examined thoroughly using a narrow fiberoptic endoscope. It is important as sinus diseases usually start in the middle meatus.
- *Transillumination*—it is performed in the darkened room by insertion of an electrically safe bright light into the mouth (with the lip closed) after removal of maxillary prosthesis. A difference in luminosity between two sinuses which are seen best at the medial third of the infraorbital rim indicates the presence of unilateral disease. A large cyst filled with clear fluid will be opaque on radiograph but brilliantly clear on transillumination. Good transillumination indicates presence of air in the sinus while failure of transillumination indicates presence of pus, solid lesion or mucosal thickening.
- *Bacteriology and cytology*—proof puncture of the antrum is usually performed through the inferior meatus to confirm radiological appearances. After topical application of vasoconstrictor and analgesic spray, a 'trocar and cannula' is passed under the attachment of the inferior turbinate up to the genu where it will naturally rest. The body of trocar is held in the palm of the hand with the index finger running along its haft to provide control and a stop. It is directed towards the tragus of the ipsilateral ear while the patient's head is held still. Moderate pressure with a rotary movement will lead to perforation of the thinnest part of the lateral inferior meatal wall, which is just behind the lacrimal duct orifice. Trocar and cannula is advanced to the lateral sinus wall and then withdrawn several millimeters before the trocar is removed. An aspirate is withdrawn into an empty syringe and sent for bacteriological and cytological examination.
- *Fiberoptic antroscopy*—in this, sinuses unresponsive to treatment or any suspicious areas which is seen radiographically, can be examined by direct vision through an endoscope. It can be performed under local

anesthesia as an outpatient procedure. The antrum is entered using a sharp trocar and cannula which can be pushed directly through the anesthetized oral or nasal mucosa, bone and sinus mucosa into the antrum. The trocar is then removed and the endoscope is inserted along the cannula and attached to a light source. It is mainly used in orbital blowout fracture where clinical and radiological examination is inconclusive.

• *Radiography:*

- *Periapical*—it will show the floor, the base of antral cavity and relationship with upper posterior teeth.
- *Caldwell posteroanterior*—good visualization of frontal sinus and ethmoidal air cells, nasal cavity and superior portion of the maxillary antrum.
- *Water's view*—visualization of maxillary sinus, when the mouth is open; sphenoid sinus can also be seen. It usually shows the roof or upper borders, medial wall and allows comparison of both maxillary sinuses.
- *Lateral skull view*—examination of sphenoid and maxillary sinuses especially anterior and posterior walls.
- *Submentovertex*—to define extent of the sphenoid sinus.
- *Computed tomography*—delineation of soft tissues in the sinus. It allows comparisons of the two sides (Fig. 27-2) of maxillary sinus.

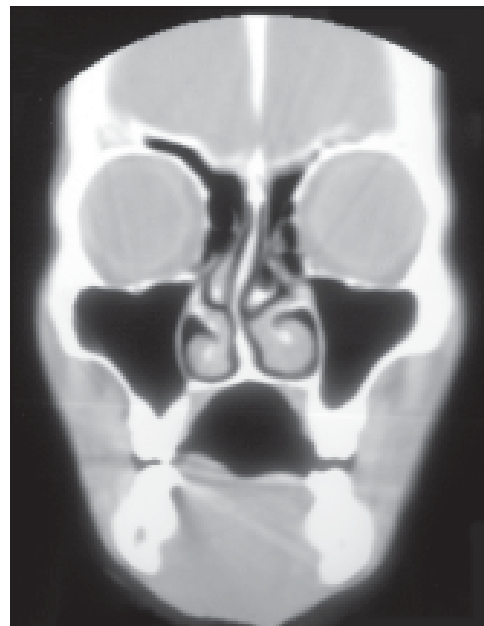


Fig. 27-2: CT scan showing maxillary sinus.

- *Scintigraphy*—radioactive isotopes are used diagnostically to demonstrate physiological changes in tissue that often precede anatomical changes or therapeutically to destroy tumor cells. Extension of antral carcinoma to involve bone produces an osteoblastic

response ahead of the advancing malignant bone destruction. This is evident in the delayed phase of a radionuclide bone scan as bone destruction becomes visible on the radiograph.

- **Ultrasound**—bony wall of the sinus is so thin that high power, short duration sound waves from a transmitter are able to pass through it. They are reflected back to the receiver when they hit an impenetrable object. It is effective in distinguishing normal sinuses, chronically inflamed sinus lining and if sinus is filled with fluid, tumor or scar.
- **Magnetic resonance imaging**—MRI is extremely sensitive in demonstrating maxillary antrum pathology due to the high signal intensity on T₂-weighted image of almost soft tissue abnormalities, contrasted with the absence of signal from both air within the sinus and the surrounding cortical bone.

Classification of Maxillary Sinus Disorders

It is described in Tables 27-1 to 27-3.

Table 27-1: First classification

A. Traumatic	
• Fracture of maxilla, nasal bone, zygoma and orbital floor • Hematoma • Foreign bodies—root piece, bullet injury, antroliths • Oroantral fistula	
B. Inflammatory	
• Acute and chronic sinusitis • Local hyperplasia from odontogenic infection • Antral polyp • Osteomyelitis	
C. Cysts	
<i>Intrinsic</i>	<i>Extrinsic</i>
• Mucus retention cyst • Serous cyst • Pseudocyst	• Odontogenic • Radicular • Dentigerous • Primordial • Keratocyst • Non-odontogenic • Globulomaxillary • Traumatic • Aneurysmal bone cyst (ABC)
D. Neoplasms	
<i>Odontogenic</i>	
• Ameloblastoma • Adenoameloblastoma • Calcifying Epithelial Odontogenic Tumor (CEOT)	
<i>Non-odontogenic</i>	
• Exostosis • Enostosis	
<i>Malignant</i>	
• Squamous cell carcinoma • Midline lethal granuloma	
E. Metabolic	
• Fibrous dysplasia • Leontiasis ossea	

Table 27-2: Second classification

A. Infection/Inflammation	
• Acute sinusitis • Chronic sinusitis	
B. Trauma	
• Oro-antral communication • Fracture of the maxillofacial skeleton • Foreign bodies within the antrum	
C. Cysts	
• Intrinsic • Extrinsic	
D. Tumors	
• Intrinsic • Extrinsic	
E. Others bone abnormalities	
• Fibrous dysplasia • Paget's disease • Osteopetrosis	

Table 27-3: Third classification

Developmental	
• Crouzon syndrome • Treacher Collins syndrome • Binder syndrome	
Inflammatory	
• Mucositis • Maxillary sinusitis • Empyema	
Cyst	
<i>Non-dental</i>	
• Benign mucosal cyst of maxillary antrum • Mucocele • Surgical ciliated cyst	
<i>Dental</i>	
• Radicular cyst • Dentigerous cyst • Globulomaxillary cyst • Odontogenic keratocyst	
<i>Benign tumor</i>	
• Antral polyp • Antral papilloma • Osteoma • Ameloblastoma	
<i>Malignant tumor</i>	
• Squamous cell carcinoma • Invasion of the maxillary sinus by local malignant disease • Metastatic carcinoma of the maxillary sinus	
<i>Traumatic</i>	
• Fractured root • Sinus contusion • Blowout fracture • Isolated injury • Complex fracture • Oroantral fistula • Foreign bodies	
<i>Calcification</i>	
• Antroliths	
<i>Miscellaneous</i>	
• Fibrous dysplasia • Pseudotumor	

Developmental Disorders

- *Crouzon syndrome*—early synostosis of the sutures produce hypoplasia of the maxillae and therefore the maxillary sinus, together with the high arched palate.
- *Treacher Collins syndrome*—Mandibulofacial dysostosis is associated with grossly and symmetrically underdeveloped maxillary sinuses and malar bones.
- *Binder syndrome*—maxillonasal dysplasia (Binder syndrome) involves hypoplasia of the middle third of the face. There is maxillary retrognathism, smaller maxillary length and frontal and maxillary sinus hypoplasia.

Inflammatory Disorders

They are caused by chemical irritation, introduction of foreign body, facial trauma, etc.

Mucositis

It is the name given when there is thickened mucous membrane of maxillary sinus. Normally, mucous membrane of maxillary sinus is about 1 mm in thickness. When the lining mucosa becomes inflamed from either infection or an allergic process, it may increase in thickness by about 10 to 15 times. This inflammation is called as *mucositis*.

Causes

- *Inflammatory disease*—dental inflammatory lesion such as periodontal disease or periapical disease may cause localized mucositis.

Clinical Features

It is usually asymptomatic and it is discovered on routine radiograph.

Radiological Features

- *Appearance*—image is detected on a radiograph as a band, noticeable more radiopaque than the air filled sinus and paralleling the bony wall of the sinus.
- *Size*—mucosal thickening can vary from a few millimeters to a thick, hypertrophic, redundant mucosa (**Fig. 27-3**).

Diagnosis

- *Clinical diagnosis*—not possible to make clinical diagnosis.
- *Radiological diagnosis*—parallel radiopaque band seen in the wall of sinus will diagnose this lesion.

Management

- *Removal of cause*—it will disappear after removing the cause.



Fig. 27-3: CT coronal section showing fine mucosal thickening, involving medial and lateral wall along with the floor of the maxillary sinus bilaterally (Courtesy Dr Ashok L).

Maxillary Sinusitis

Inflammation of the mucosa of the paranasal sinuses is referred to as *sinusitis*. When maxillary sinus is involved, it is called as *maxillary sinusitis*. When all the sinuses are involved, it is called as *pansinusitis*.

Types (depending upon duration)

- *Acute*—This type is associated with severe pain in sinus area.
- *Subacute*—it is an interim stage between acute and chronic sinusitis.
- *Chronic*—it develops as a result of neglected and overlooked dental focus of infection. The lining becomes thicker and irregular.

Etiology

Dental causes

- *Periapical infection from the teeth*—it may follow dental infection particularly from upper molar and premolar teeth. The anatomic proximity of the roots of the maxillary bicuspid and molar teeth to the floor of the sinus leads to potential infection of the sinus by direct extension of an apical abscess.
- *Oroantral fistula*—the accidental opening in the floor of the maxillary sinus during dental extraction is called as oroantral opening.

- *Periodontitis*—it may spread from a deep pocket of marginal periodontitis.
- *Traumatic*—injury of facial bones especially nasal bones and malar bones.
- *Dental material in the antrum*—perforation of endodontic filling substance. If root canal is overfilled then there are more chances of gutta purcha point to be inserted into the maxillary sinus.
- *Implant*—implants are used in the upper edentulous jaw to aid the retention of dentures or bridges or replace missing teeth. Implants are also used when there is insufficiency of bone to support the denture. In these cases as bone is thin, implant can penetrate the nose or sinus.
- *Infected dental cyst*—cysts which have become infected and involve the maxillary sinus can also cause sinusitis.

Non-dental causes

- *Mechanical obstruction of ostium*
 - *Common cold*—it is the most common cause of mechanical obstruction of ostium. It will produce inflammatory edema of the nasal mucosa which obstruct the antronasal duct and cause mucus to accumulate in the sinus. Trapped mucus becomes secondarily infected by local commensal bacteria.
 - *Allergic rhinitis*—it may cause maxillary discomfort due to edema round the ostium and retention of secretion.
 - *Other conditions*—deviated nasal septum, presence of nasal polyp and prolonged nasotracheal intubation can cause stagnation and relative anaerobiasis.
 - *Direct bacterial contamination*—infected material may also be introduced directly by jumping or hydroskiding feet first into contaminated water without holding the nose or during diving, when pressure changes in the nose force nasal secretion into the sinus.
 - *Immune deficiency*—sinusitis can occur in immune deficiency state, like leukemia, lymphoma and AIDS.
 - *Influenza*—it can also occur in influenza when bacteria invade as secondary microorganisms.
 - *Blood borne infection*—it can also occur in some cases of blood borne infection.
 - *Disease of maxillary sinus*—benign mucosal cyst or tumors of the maxillary sinus can also lead to maxillary sinusitis.
- increased by biting on the affected side but unaffected by drinking hot, cold or sweet fluids. Pain may be referred to various areas including the teeth, orbit, head and ear. Pain in the teeth may be referred to the premolars and molars on the affected side. Teeth in the involved side become sore and painful.
- *Postnasal drip*—postnasal drip may cause irritation, stuffiness and nasal discharge. Due to this, patient may get difficulty in breathing.
 - *Generalized toxemia*—generalized toxemia develops, i.e. fever with chills, dizziness, malaise and nausea.
 - *Nasal discharge*—nasal discharge is watery in the beginning but later becomes mucopurulent. In cases of sinusitis from infected teeth, the discharge has a foul odor.
 - *Nasal mucosa*—the nasal mucosa of the anterior nares may show reddening and inflammation and there may be presence of pus.
 - *Tenderness on palpation*—tenderness to pressure or swelling over the involved sinus. Since the anterolateral and posterolateral sinus walls are thinnest in the area above the tooth root, thumb pressure on the cheek region is the best way to elicit tenderness externally. Premolar and molars of the affected side may be sensitive to percussion.
 - *Intraoral finding*—intraorally, there may be a mucobuccal swelling, reddening and pain near the sinus region.
 - *Postural test*—if no pus is seen in the middle meatus, it is decongested with a pledge of cotton soaked with a vasoconstrictor and patient is made to sit with the affected sinus turned up. Examination after 10-15 minutes may show discharge in the middle meatus.
 - *Complication*—the most common complication is spread of infection to the orbit. Orbital swelling, pain and loss of vision can also occur.

Subacute

- *Symptoms*—It is devoid of symptoms associated with acute congestion such as pain and generalized toxemia.
- *Discharge*—discharge is usually purulent and associated with a nasal voice and stuffiness.
- *Soreness*—soreness of throat is common feature.
- *Disturbed sleep*—patient can not sleep well due to cough that irritates the patient constantly.

Chronic

- *Symptoms*—general symptoms of chronic sinusitis include sense of tiredness, low grade fever and feeling of being unwell. Stuffy sensation over the affected side of the face.
- *Other features*—there is nasal obstruction, nasal discharge and headache are the related symptoms.

Clinical Features

Acute maxillary sinusitis

- *Symptoms*—the main symptom is severe pain which is constant and localized. Pain may be felt in the area of eyeball, cheek and frontal region. Pain may be exacerbated by stooping or lowering the head. Pain is

Radiographic Features

- **Radiodensity**—radiographically, the thickening of the mucous membrane and the accumulation of secretions that accompany sinusitis reduce the air content and it will appear as radiopaque (Fig. 27-4).

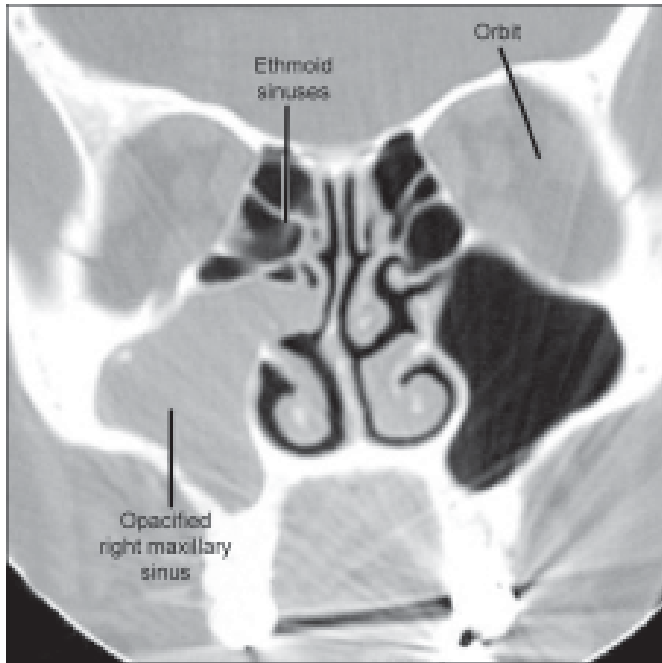


Fig. 27-4: Opacification seen in right maxillary sinus due to sinusitis (Courtesy Dr Ashok L).

- **Four patterns are seen:**
 - **Localized thickening**—localized thickening at the base of the sinus.
 - **Generalized thickening**—roughly generalized thickening of mucoperiosteum around the entire wall of the sinus.
 - **Complete filling except ostium**—complete filling of the sinus except in the region of the ostium on the medial wall.
 - **Complete filling**—complete filling of the sinus.
- **Allergic sinusitis**—in the cases of allergy, mucosa will be more lobulated in contrast to that in infection where it is straighter and parallel to the sinus wall.
- **Chronic sinusitis**—chronic sinusitis may result in persistent opacification of the sinus and sclerosis or thickening of surrounding bone.
- **Antral halo appearance**—Sometimes if infected teeth are involved then inflammatory changes may lead to resorption of the antral floor and remodeling to produce the appearance described as an antral halo.
- **Resolution of sinusitis**—resolution of acute sinusitis will appear as small clear areas appear in the interior of the sinus as the thickened mucosa gradually shrinks.

Diagnosis

- **Transillumination test**—affected sinus will be found opaque.
- **Radiograph**—Water's view (Fig. 27-5) and OPG can be taken.



Fig. 27-5: Water's view showing complete opacification of left maxillary sinus (Courtesy Dr Ashok L).

- **Laboratory diagnosis**—there is elevated leukocyte count. Lining of maxillary sinus may show a typical acute inflammatory infiltrate with edema of the connective tissue and often hemorrhage. In chronic cases, cellular proliferation is present.

Management

- **Removal of the cause of dental infection**—treatment should be directed to drain the periapical abscess from the root canal or by extracting the teeth.
- **Antibiotic**—most commonly used antibiotic is doxycycline hydrochloride. Doxycycline is given in the dose of 100 mg daily. Penicillin is also the drug of choice as it is effective against most of the anaerobic microorganisms. In some cases, amoxicillin or clotrimazole can be given.
- **Analgesic and anti-inflammatory drugs**—these drug are used to control pain and inflammation. Most commonly used are Ibuprofen or Nimesulide.
- **Nasal decongestant**—Ephedrine nasal drops (0.5%) are most commonly used and give relief for several hours. Xylometazoline (0.1%) is the alternative drugs to Ephedrine but it can lead to more rebound effect.

- *Stem inhalation*—it may act by hydrating the mucous layer, making it less viscous and thereby encouraging normal ciliary clearance of the maxillary sinus. The use of volatile aromatic additive, such as menthol or eucalyptus can be used to stimulate cold receptor nerve endings.
- *Antral lavage*—if antibiotics and nasal drops fail to resolve the condition, pus may be removed from the antrum. This procedure is carried out under local anesthesia by inserting hollow trochar and cannula into the antrum through the nasal wall beneath the inferior turbinate. Warmed saline is syringed through the cannula so that the fluid and pus return via the antranasal duct.
- *Antrostomy*—if all the above procedures fail to cure the sinusitis and infection is recurrent then inferior meatal antrostomy and middle meatal antrostomy can be done.
- *Transnasal endoscopic surgery*—endoscopic approach is made to the middle meatus using rigid instrument to visualize the infundibulum and the natural opening of the sinuses.

Cysts

Cysts are classified into dental and nondental.

Nondental Cyst

Nondental cysts of maxillary sinus are as follows.

Antral Pseudocyst or Benign Mucosal Cyst of Maxillary Antrum

It is also called as, '*retention cyst*', '*pseudocyst*', '*lymphangiectatic cyst*', '*false cyst*', '*benign mucosal antral cyst*', '*serous non-secretory retention pseudocyst*', '*mesothelial cyst*' and '*intramural cyst*'.

Pathogenesis

- *Inflammatory exudate*—severe inflammation around the ducts of mucus glands of the antral lining may alter their integrity. When the patient sneezes, mucus can be expelled into soft tissue through the wall of such injured duct. Once the pathway for extraglandular accumulation of mucus has been established, the process may continue until a cyst has developed. Infection may result from odontogenic source or it may cause due to allergy.
- *Mechanical factors*—because of frequent location of the lesion at the sharp angle between the floor and frontal or lateral aspect of the sinus cavity close to the alveolar process mechanical factors might be involved in the development of cyst. Mucosal swelling, perhaps the result of a previous common cold may lead to mechanical stress leading to the rupture of sharp angle tissue or its bony attachment.
- *Proteolytic process*—some have also found a low concentration of alpha-antitrypsin in the cystic fluid which suggests the possibility that a proteolytic process could lead to the expansion of the cyst.

Clinical Features

- *Age and sex*—the great majority of cases were found in patients in the age group of 21-30 years. Males are more commonly affected than females in the ratio of 2:1.
- *Sites*—most commonly involved sites are the antral floor and the lateral wall of maxillary sinus.
- *Symptoms*—there may be localized dull pain in the antral region, or fullness and numbness of the cheek. If it is completely filled sinus, it will prolapse through the ostium and cause obstruction and postnasal drip. There may be pain in the teeth and face over or near the sinus. There may be stuffiness, fullness, headache, symptoms of cold, and numbness of upper lip.
- *Signs*—sometimes, antral swelling may also occur. There is copious discharge of yellow fluid from the nostrils. Retention of pseudocyst may enlarge and fill the sinus cavity completely, it frequently ruptures as a result of abrupt pressure change caused by sneezing or blowing the nose. Expanding cyst will cause herniation through the ostium into the nasal cavity where it subsequently ruptures.

Radiological Features

- *Radiodensity*—it is homogenous mass that is more radiopaque than the surrounding sinus cavity.
- *Appearance*—It appears as a soft tissue mass rather than a calcified area so that medial and lateral landmarks can generally be visualized through the lesion.
- *Site*—It is found projecting from the floor of the sinus, although some may form on the lateral walls.
- *Shape*—the cyst appears as a spherical, ovoid or dome shaped radiopacity (**Fig. 27-6**).
- *Margins*—it has a smooth and uniform outline. The smooth curved borders are well defined but not well corticated.
- *Base*—they may have a narrow or broad base.
- *Size*—they vary in size from minute to very large and may occasionally occupy the entire maxillary sinus. Image may be of a fingertip to that of a completely filled sinus making it opaque.
- *Bone*—there is no resorption of adjacent bone and of particular importance is the persistence of thin radiopaque line of the antral cortex.



Fig. 27-6: Dome shaped radiolucency seen in left maxillary sinus suggestive of antral pseudocyst.

- Mucus type is associated with thickened mucosa while serous type appears normal.

Diagnosis

- *Clinical diagnosis*—dull pain in antral area, discharge of yellow fluid from nostril should suspect this cyst.
- *Radiological diagnosis*—ovoid and dome shaped radiopacity projecting from the floor will diagnose this condition.
- *Laboratory diagnosis*—biopsy shows infiltration of chronic inflammatory cells in connective tissue wall of the cyst. Cystic fluid is thick, tenacious, white translucent and sterile.

Differential Diagnosis

- *Inflammatory lesions of the sinus*—in inflammatory lesions, patient may complain of pain which is absent in case of cyst.
- *Apical radicular cyst*—tooth is not vital.
- *Odontogenic cyst*—mucus retention cyst is dome shaped and does not have thin marginal line representing the hyperostotic borders characterized by odontogenic cyst. Odontogenic cysts are more rounded and tear shaped and most are not so homogenous, as mucus retention cyst. Lamina dura is not intact in odontogenic cysts.
- *Antral polyp*—they are more opaque, more heterogeneous and multiple commonly associated with thickened mucosa.
- *Surgical ciliated cyst*—more rounded and more radiolucent.

- *Neoplasm*—the neoplasm is less likely to be dome shaped as the retention cyst.

Management

- *Removal of foci of infection*—the adjacent tooth should be treated for any source of infection.
- *Caldwell-Luc operation*—large cysts should be removed through Caldwell-Luc operation.

Sinus Mucocele

It is an expanding destructive lesion that begins with the development of mucus retention cyst in a blocked ostium. If mucocele becomes infected, it is called as pyocele or mucopyocele.

Etiopathogenesis

- *Obstruction of sinus ostium*—blockage or obstruction may result from long and tortuous infundibulum, intranasal or intra-antral inflammation, and from polyps and bony neoplasm. This blocked ostium will act as separate cyst like structure lined by epithelium and filled with mucin.
- *Accumulation of mucin*—the cystic lesion continues to accumulate mucus. After it has filled the sinus cavity the pressure increases and the lesion will become destructive by expanding and destroying sinus wall by thinning out the bone wall.

Clinical Features

- *Site*—there is predilection for ethmoid and frontal sinuses due to the relative difficulty as a cyst has protruding through the longer and narrower nasofrontal duct and infundibulum into the nasal cavity, in contrast to the shorter and larger ostium in case of maxillary and sphenoidal sinuses.
- *Symptoms*—it may exert pressure on the superior alveolar nerve in the resorbing sinus wall and cause radiating pain. Sensation of fullness in the cheek may be accompanied swelling over the anteroinferior aspect of the antrum, the area where the wall is thinnest. If the cyst expands inferiorly, it may cause loosening of the posterior teeth in that area. If it expands medially, lateral wall of the nasal cavity is deformed and nasal airway is obstructed.
- *Signs*—if expands towards the orbit, it may cause diplopia and proptosis (protrusion of the globe of the eye).

Radiographic Features

- *Site*—about 90% of the cases are in ethmoidal and frontal sinuses and are rare in maxillary and sphenoid sinuses.

- **Radiodensity**—there is opacification of sinus.
- **Shape**—the shape of the sinus is changed into a more circular shape as the mucocele grows.
- **Effect on surrounding structures**—the shape of sinus changes with bony expansion. There is erosion of septa and the bony wall may occur. Borders of the expanding sinus become sclerotic.
- **Frontal sinus**—scalloped borders of frontal sinus become smooth by erosion and intersinus septum may be displaced.
- **Maxillary sinus**—teeth may be displaced or resorbed.
- **Ethmoid sinus**—displacement of the lamina papyracea may occur, displacing the content of the orbit.
- **Sphenoid sinus**—the expansion may be in a superior direction, suggesting a pituitary neoplasm.
- **Orbit**—Bone of supramedial border of the orbit is destroyed or displaced.

Diagnosis

- **Clinical diagnosis**—symptoms of sinus mucocele are severe and consist of radiating pain, diplopia and obstruction of nasal airway.
- **Radiological diagnosis**—opacification of sinus with erosion of septa. Displacement of teeth can also occur.

Differential Diagnosis

- **Malignancy**—any suggestion of a lesion associated with occluded ostium should be mucocele.

Management

- **Surgical**—surgical removal by the Caldwell-Luc operation is traditional method for management of sinus mucocele.
- **Endoscopic middle meatal antrostomy**—nowadays, this treatment module gives good results.

Postoperative Maxillary Cyst

This is also type of sinus mucocele which occurs after trauma. It is also called as '*surgical ciliated cyst of maxilla*'. Its name arises from the fact that it has a delayed complication arising within years after surgery involving the maxilla.

Pathogenesis

- **Trapping of epithelium in wound closure**—they are derived from the epithelial lining of the maxillary sinus which is trapped in the wound during closure of the Caldwell-Luc incision and subsequently begins to proliferate. Sinus lining will separate from the sinus and epithelium lined cavity is formed in which mucin is secreted. This type of cyst is originated after difficult extraction in which sinus lining is damaged.

Clinical Features

- **Age and sex distribution**—the great majority of the patients are in the fourth and fifth decades and their age ranges from 21 to 72 years. There is a preponderance of males to females (2:1).
- **Symptoms**—pain, discomfort or swelling in the cheek or face or intraorally in the palate or alveolus is a common complaint.
- **Sign**—pus may be discharged.

Radiological Features

- **Site**—well defined radiolucency closely related to the maxillary sinus.
- **Margin**—surrounding bone sclerosis is evident in at least a part of the bony margin. There is pneumatization and relatively well defined unilocular radiolucent margins showing marked osteosclerotic changes.
- **Pressure effect**—occasionally, cyst appears to encroach on the sinus itself but lack of communication between the two has been demonstrated by injecting the sinus with a radio-opaque material. As the lesion enlarges, sinus walls become thinned and eventually perforate and may resemble a malignant neoplasm. Resorption of maxillary alveolar processes also occurs.

Diagnosis

- **Clinical diagnosis**—history of trauma or operation followed by pain and discomfort in the sinus region.
- **Radiological features**—contrast study will help to diagnose this cyst. Well defined radiolucency close to sinus will give clue to diagnose the case.
- **Laboratory diagnosis**—cysts are lined by pseudo-stratified columnar epithelium with squamous metaplasia in chronically inflamed cases. Combination of ciliated, cuboidal and squamous epithelium with varying numbers of mucus cells may be seen.

Management

- **Enucleation**—enucleation through an approach appropriate to the site is the treatment of choice. Recurrence may occur if the wall is very thin and there is perforation of the bone making enucleation difficult.

Dental Cyst

These are described in detail in Chapter 13: Cysts of Jaw.

- **Radicular**—those that develop in the maxilla may extend into the maxillary sinus. They may cause elevation of the floor of the sinus resulting in a halo appearance. Large cysts may obliterate the sinus and make differentiation of this form from sinusitis is difficult.

- *Dentigerous cyst*—when they expand into the sinus, the radiograph shows radiolucency elevating an intact wall or floor of the maxillary sinus. If it is small, it is usually round, dome shaped opacity in the base of the antrum with well defined radiopaque corticated margins to the edge of the meniscus. Sometimes, there may be displacement of the associated tooth (**Fig. 27-7**). If the cyst is large then there is total opacity of the antral region due to complete compression of the antral cavity. There is also a loss of antral outline.
- *Cyst in the globulomaxillary area*—they may expand so as to obliterate or alter the typical pattern or border of nasal fossa and anterior process of the maxillary sinus.

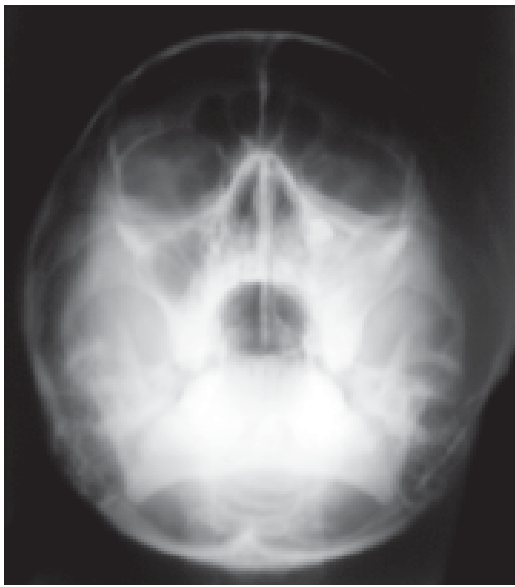


Fig. 27-7: Dentigerous cyst involving maxillary sinus. Note the displacement of the tooth in the sinus.

Benign Tumor

Antral Polyp

The thickened mucosa of chronically inflamed sinus frequently form irregular folds called as '*polyps*'. Polypoid atrophy of mucosa may develop into an isolated area or number of areas throughout the sinus.

Clinical Features

- *Age*—it usually occurs in young persons.
- *Site*—maxillary sinus is more involved as compared to other sinus. In maxillary sinus they may arise from any part of the sinus wall and occasionally pass through the ostium to appear in the nose as antrochoanal polyps.
- *Symptoms*—patients present with nasal obstruction, pain is very mild on pressure as mass present inside the nose.
- *Saints triad*—it is associated with "Saints triad", i.e. nasal and antral polyposis, aspirin sensitivity and asthma.
- *Exacerbation of asthma*—polyps may exacerbate the asthma by causing obstruction of the nose. It is most commonly pedunculated, or sessile mass which grows very slowly. After the polyp grows to occupy most of the antrum it frequently herniates into the nasal cavity. This may be brought about by repeated sneezing or nose blowing in about 4-6% cases.

Radiological Features

- *Appearance*—it appears as homogenous soft masses with smooth, outwardly convex borders. Single or multiple lesions may be present. If polyp occurs in the roof of the maxillary sinus in a patient with a history of trauma, the plain film examination may simulate a blow out fracture.
- *Destruction of walls of sinus*—Polyps may cause destruction or displacement of bone. They can displace or destroy medial or lateral wall.
- *CT features*—have mucoid attenuation with mucosal enhancement seen at the polyps' surface. It appears as smooth homogenous mass.
- *MRI features*—polyps will have low to intermediate T₁-weighted and high T₂-weighted signal intensities. Mucosa adjacent to polyps will enhance as compared (**Fig. 27-8**) to polyps.

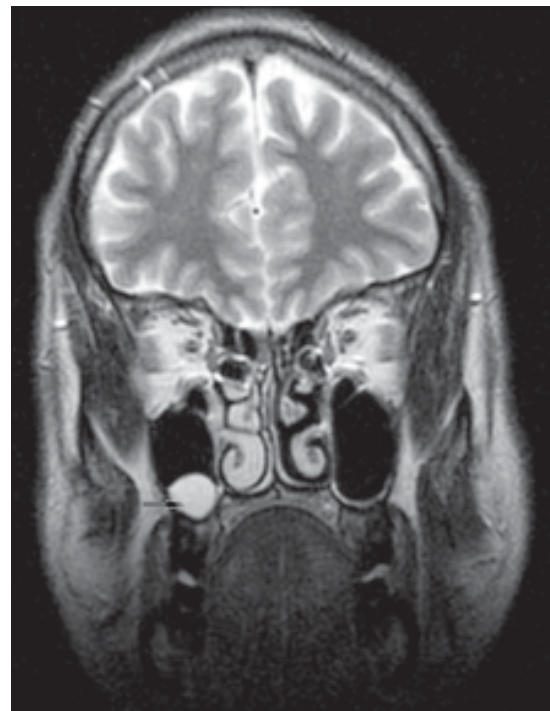


Fig. 27-8: MRI T₂ coronal section image showing well margined hyperintense area in the floor of the right maxillary sinus (Arrow mark) suggestive of solitary polyp (Courtesy Dr Ashok L).



Fig. 27-9: Polyp seen in left maxillary sinus as homogenous radiopacity (Courtesy Dr Bhaskar Patle).

Diagnosis

- *Clinical diagnosis*—saint tried with pain and pressure inside the nasal cavity may give clue to the diagnosis.
- *Radiological diagnosis*—CT and MRI will diagnose the polyp as homogenous radiopaque mass in the sinus (Fig. 27-9).

Management

- *Excision*—simple excision is close with endoscopic surgery or Caldwell-Luc operation.

Antral Papilloma

It is a rare tumor of respiratory epithelium that occurs in the nasal cavity and the paranasal sinuses.

Clinical Features

- *Age and sex*—it is seen in the age group of 20 to 25 years with predominance in males.
- *Site*—usually ethmoid and maxillary sinuses are involved.
- *Symptoms*—there is unilateral nasal obstruction, nasal discharge, pain and epistaxis can occur. Recurring sinusitis and subsequent nasal obstruction on the same side.
- *Appearance*—it has got warty appearance.

Radiographic Features

- *Site*—it appears as isolated polyps in the nose and sinus.
- *Radiodensity*—the neoplasm appears a homogenous radiopaque mass of soft tissue density (Fig. 27-10).
- *Pressure effect*—bone destruction can occur due to pressure erosion.

Diagnosis

- *Clinical diagnosis*—warty appearance with recurring sinusitis may suspect this disease.



Fig. 27-10: Homogeneous radiopaque mass with destruction of wall of right maxillary sinus seen due to papilloma.

- *Radiological diagnosis*—homogenous mass with bone destruction will give clue to the diagnosis.

Management

- *Excision*—surgical excision of tumor should be carried out.

Osteoma

It is most common mesenchymal neoplasm in the paranasal sinus.

Clinical Features

- *Age and sex*—it is more common in 2nd, 3rd and 4th decade. It is more common in male as compared to females and the ratio is 2:1.
- *Symptoms*—it is slow growing and asymptomatic. When symptoms occur, they are as a result of obstruction of the sinus ostium or infundibulum or are secondary to erosion or deformity, orbital involvement or intracranial extension. It may extend in the nose and cause nasal obstruction or swelling on the side of the cheek.
- *Signs*—it may expand into the sinus and produce swelling of the cheek or hard palate. In cases of extension to orbit the patient may have proptosis.

Radiological Features

- *Site*—they occur more often in frontal and ethmoidal sinuses. The maxillary sinuses are also involved.
- *Shape*—lobulated or rounded homogenous masses of high density are seen.
- *Margins*—they have sharply defined margins.

Diagnosis

- *Clinical diagnosis*—nasal obstruction, proptosis may give some clue to the diagnosis.

- *Radiological features*—lobulated homogenous mass of high density is present.

Management

- *Excision*—this tumor should be surgically removed.

Ameloblastoma

It is most common extrinsic tumor affecting the maxillary sinus. It may cause loosening of teeth, nasal obstruction and painless facial deformity. Sinus cavity is invaded at an early stage. There is radiographic appearance equivalent to soft tissue density. Antral cavity is expanded and filled with soft tissue mass. Bony wall is eroded.

Malignant Tumor

Squamous Cell Carcinoma

It originates from metaplastic epithelium of the sinus mucous membrane lining. It is the most common primary tumor of paranasal sinuses comprising 80 to 90% of cancers in this site.

Etiology

- *Idiopathic*—Most cases of squamous cell carcinoma of maxillary sinus are without any cause.
- *Sinusitis*—respiratory epithelium is known to undergo squamous metaplasia in the presence of infection and chronic sinusitis can be predisposing factor for antral carcinoma.
- *Snuff and smoke*—the use of indigenous snuff and the smoky atmosphere may be causal factor for carcinoma of paranasal sinus.
- *Occupational hazards*—it is more common in boot and shoe and nickel worker. Adenocarcinoma of the nasal passage is an occupational hazard for furniture workers.

Clinical Features

- *Age and sex*—mean age of occurrence is 60 years. Males are commonly affected more than females in the ratio of 2:1.
- *Symptoms*—there is facial pain, swelling, nasal obstruction. Patient also complains unilateral nasal stiffness. When the second division of trigeminal nerve is involved, there is intense pain or paresthesia of midface occurs.
- *Signs*—there is ulceration or mass seen on the hard palate (Fig. 27-11).
- *Lymph nodes*—lymphadenopathy is always present in carcinoma of maxillary sinus.
- *Medial wall involvement*—medial wall involvement leads to nasal obstruction, discharge, bleeding and pain.



Fig. 27-11: Ulceration seen on first molar area due to maxillary sinus malignancy.

Epiphora will result if the lacrimal sac or nasolacrimal duct is obstructed.

- *Floor involvement*—involvement of the floor of the sinus leads to expansion of the alveolus, unexplained pain, numbness of teeth, loosening of teeth and swelling of the palate or alveolar ridge and malfitting dentures. It may erode the floor and penetrate the oral cavity (Fig. 27-12).
- *Lateral wall involvement*—lateral wall involvement leads to facial and vestibular swelling, pain and hyperesthesia of maxillary teeth (Fig. 27-13).
- *Roof involvement*—roof involvement leads to diplopia, proptosis and pain over the cheek and upper teeth (Fig. 27-14).
- *Posterior wall involvement*—posterior wall involvement leads to painful trismus, obstruction of Eustachian tube causing stuffy ear, referred pain and hyperesthesia over the distribution of second and third division of trigeminal nerve.



Fig. 27-12: Ulceration and perforation seen in hard palate in maxillary sinus malignancy (Courtesy Dr Ashok L).



Fig. 27-13: Swelling in vestibular region occur due to involvement of lateral wall (Courtesy Dr Ashok L).



Fig. 27-14: Involvement of roof causes swelling in infraorbital region.

- *Nerve involvement*—it may involve infraorbital nerve and produces paresthesia of the cheek or erodes blood vessels giving rise to epistaxis. Paresthesia of mandibular nerve can also occur if the tumor invades the cranium.

Radiographic Features

Small early lesion

- *Shape and radiodensity*—non-specific well defined round soft tissue opacity within the antrum (**Fig. 27-15**).
- *Antral wall*—variable destruction of the bony antral wall. Loss of fine linear outline of the lateral wall is a particularly sensitive sign of bone destruction.

Large well established lesion

- *Pressure effect*—destructive outline of the sinus destroying bone and causing irregular bony radiolucency with



Fig. 27-15: Radiopacity seen on right side of maxillary antrum which is obscuring the wall of maxillary sinus.

erosion of the medial wall. There may be destruction of the floor and anterior or posterior walls (**Fig. 27-16**).



Fig. 27-16: Destruction of walls of maxillary sinus seen in malignancy.

- *Teeth*—occasional resorption and displacement of the teeth. There may be bone destruction around the teeth or irregular widening of the periodontal ligament space.
- *Zygomatic arch*—advanced cases involve destruction of the zygomatic arch.
- *Tomography*—on the tomography, there is destruction of the surrounding hard and soft tissue (**Fig. 27-17**).

Diagnosis

- *Clinical and radiological diagnosis*—ulceration in palate with radiopacity in maxillary antrum may suggest maxillary antrum malignancy.

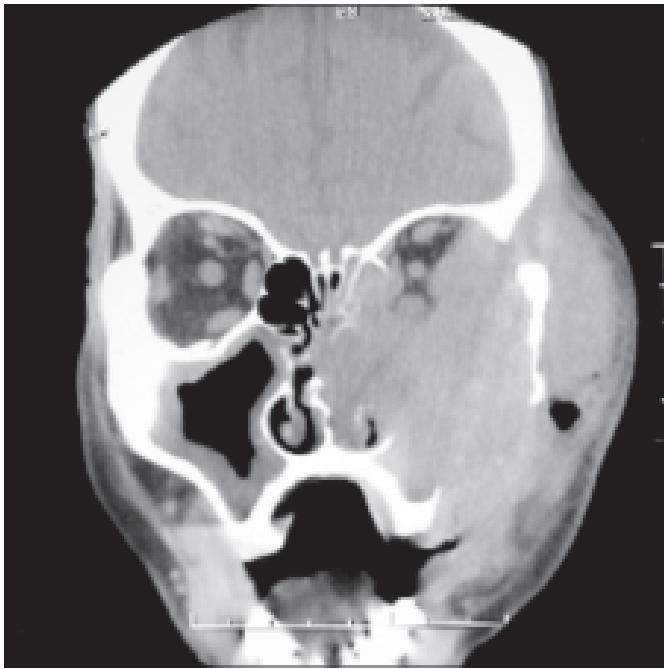


Fig. 27-17: Destruction of walls of maxillary antrum on left side seen in patient of maxillary antrum malignancy (Courtesy Dr Ashok L).

- *Laboratory diagnosis*—biopsy features are suggestive of squamous cell carcinoma.

Management

- *Cytotoxic drugs*—local intra-arterial infusion of cytotoxic drugs may be helpful for pain control.
- *Radiotherapy*—it is the main mode of treatment. A course of high voltage radiotherapy or gamma rays are given.
- *Surgery*—if radiotherapy cannot control the disease upto the expectation, excision of the maxilla should be performed.
- *Reconstruction*—after surgery, sophisticated prosthesis should be constructed.

Invasion of the Maxillary Sinus by Local Malignant Disease

Tumors of the upper jaw spread easily into the sinus. Pleomorphic adenoma arising in palatal minor salivary gland may bulge into the sinus floor and adenoid cystic carcinoma may invade it.

Metastatic Carcinoma of the Maxillary Sinus

It is rare site for the secondary tumor deposits. The most common site for the primary disease is kidney followed by the breast in females and the testicle (seminoma) in males.

Traumatic Injuries to the Paranasal Sinuses

Root in Antrum

- *Cause*—in maxillary posterior teeth, it is possible that fractured root tip may be forced into the maxillary sinus either while extraction or while removing the root tip. Following incomplete extraction of tooth, the apical segment remaining in the socket may be dislodged by injudicious use of elevators.
- *How to differentiate between root in socket and root in sinus*—if root tip is in the socket and superimposed over the sinus, then lamina dura will be intact and if it is in maxillary sinus, lamina dura will be lost.
- *Perforation of sinus*—when root tip is in the sinus and trapped under mucoperiosteum then it will be fixed and it may cause movement when it perforates the sinus.
- *Removal of root tip*—removal of the root tip can be done through the tooth socket or through the canine fossa by Caldwell-Luc approach.

Foreign Bodies

- *Root fragment and teeth*—displaced root fragments or teeth may present in the sinus.
- *Excess root canal filling*—excess root canal filling material forced through the apex of an upper posterior tooth during endodontic treatment.
- *Oroantral communication*—foreign material pushed into the antrum through an existing oroantral communication.
- *Metallic object*—metallic object such as pellets, bullets and fragments of shells or bombs.
- *Radiological detection*—the presence, position and often the nature of the foreign body can be seen by using the radiograph (**Fig. 27-18**). When roots are present in the socket, it usually reveals root canal but in some cases, there is only root tip present with no root canal seen. If you want to determine it is in the socket or in the sinus then you have to look for the lamina dura and periodontal ligament. If lamina dura and periodontal ligament is present then the root is in the socket.

Sinus Contusion

- *Cause*—blow to the face produces damage to the lining of paranasal sinuses without fracturing the facial bone. Traumatic force can be transmitted through the bony wall and be absorbed by mucosal lining which suffers damage.

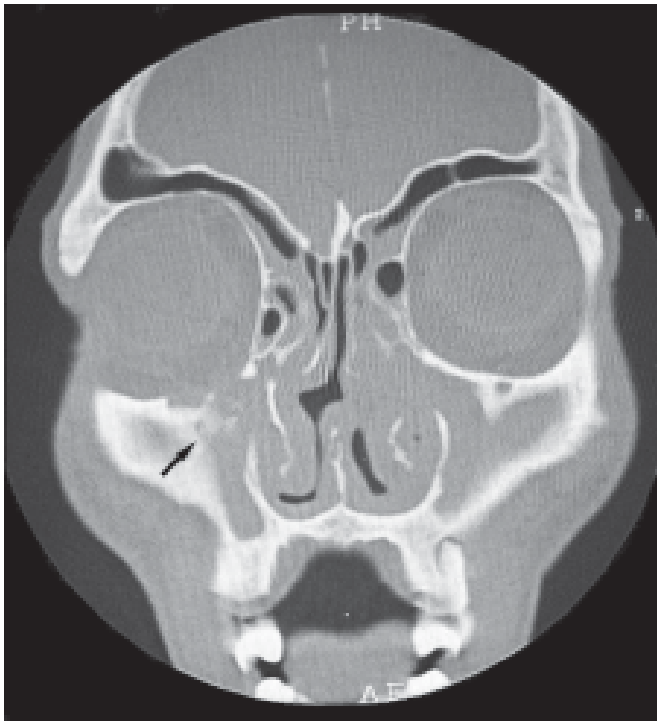


Fig. 27-18: Foreign body seen in maxillary sinus.

- *Green stick fracture*—it may cause green stick fracture of the sinus with tearing injury to the mucosal lining.
- *Radiological features*—hazy sinus is produced by edema of the mucosa, soft tissue mass that mimics a retention cyst and as a result of intramucosal hematoma, an opaque sinus or fluid level resulting from hemorrhage on mucosal tear.
- *Difference between sinusitis and sinus contusion*—in sinus contusion, there is bloody nasal discharge, extreme tenderness of involved sinus on pressure, rapid resolution of soft tissue changes.

Blow-out Fracture

- *Cause*—blow-out fracture results from sudden increase in intraorbital pressure due to blow to the eye. The pressure of blow forces the inferior orbital content (fat and muscle) through the fracture.
- *Symptoms*—diplopia (double vision) when victim looks upward (caused by entrapment of inferior rectus) and enophthalmos (backwardly depressed eyelid) following reduction of edema (Fig. 27-19) and fat atrophy.
- *Radiological features*—opacification of sinus with or without a fluid level. Soft tissue mass in upper portion of sinus and image of depressed bone fragment (orbital floor in sinus) (Fig. 27-20). It will produce a tear-drop shaped opacity in the upper part of the antrum, the 'hanging drop appearance' caused by herniation of the



Fig. 27-19: Blow-out fracture showing edema below the eye.

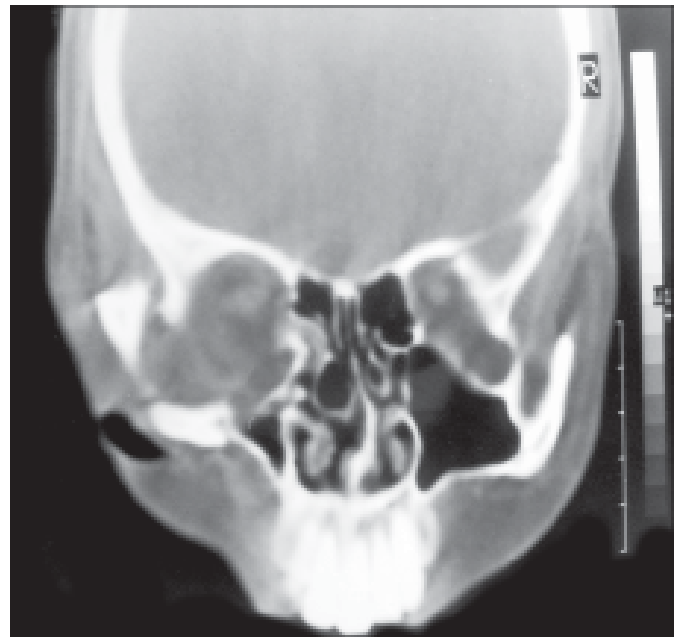


Fig. 27-20: Blow-out fracture seen on the CT.

orbital content downwards into the antrum following collapse of the antral roof. The depression fracture of the orbit is accompanied by fracture of the antrum wall of the maxillary sinus.

Isolated Fracture

It involves single wall, which are identified on the radiograph by a bright line. Anterolateral wall of the maxillary sinus is the most common site. Sometimes fracture of the floor of the maxillary sinus occurs into which roots of the posterior teeth are projecting which occurs during tooth extraction.

Zygomatic Complex Fracture

The fracture occurs at the line of weakness and passes through the orbital floor, usually medial to the zygomaticomaxillary suture and therefore inevitably involves the maxillary sinus. Fractured zygoma is forced into the sinus. It results in tearing of the lining membrane and subsequent bleeding into the antrum. Antrum will appear cloudy or show a fluid level (Fig. 27-21).

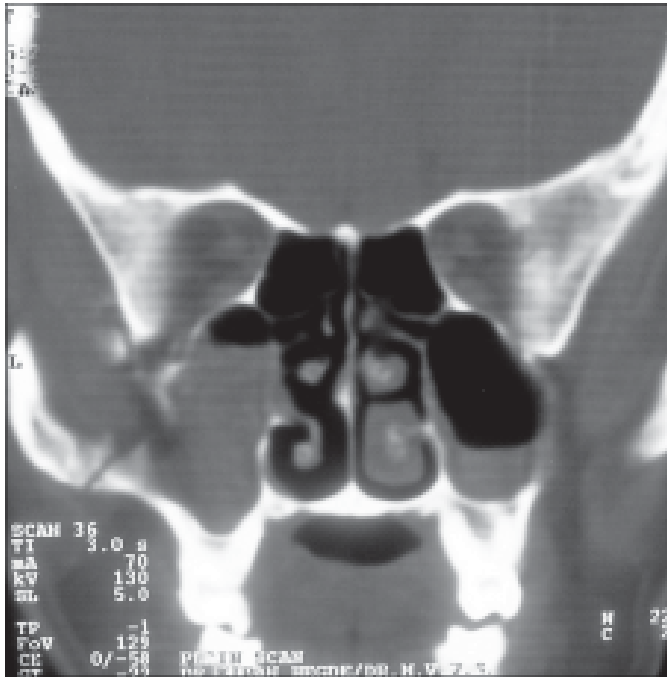


Fig. 27-21: Zygomatic complex fracture resulting in the haziness seen in the sinus.

Fractured Tuberosity

Tuberosity fracture inevitably involves the maxillary sinus. It occurs most frequently when extracting a lone standing upper third molar. If the bone is found to be moving with tooth during extraction the operator should stop immediately. Antibiotics, nasal drops and inhalation are therefore prescribed to help and prevent development of a chronic oroantral fistula.

Oroantral Fistula

It is a pathological pathway connecting oral cavity and maxillary sinus. If there is no pathological lining present, it is called as oroantral opening which is seen after tooth extraction.

Causes

- *Extraction of teeth*—teeth which is having periapical infection. Inappropriate or incorrect use of elevators during root or tooth removal may lead to oroantral fistula.

Apices of upper posterior teeth are within 3 mm of the cortical floor of the maxillary sinus or sometimes they may be inside the maxillary sinus. Forceps extraction of a solitary isolated premolar or molar in an edentulous part of the arch is also prone to cause disruption of the sinus floor.

- *Blind instrumentation*—blind instrumentation without adequate surgical exposure, in an attempted retrieval of retained apices in the upper posterior quadrant can cause oroantral fistula.
- *Surgery*—oroantral opening can occur in cases of removal of cysts and benign tumors in upper posterior region.
- *Facial trauma*—oroantral fistula may occurs following massive trauma to the middle third of the facial skeleton, especially if the face is struck by missile or sharp object which is driven through the mouth into the maxillary sinus.
- *Malignant tumor*—malignant tumors of the maxillary sinus may penetrate the lateral bony wall of maxillary sinus or it may erode through the floor of the sinus into the mouth producing oroantral communication.
- *Osteomyelitis*—it is rare, unless there is underlying systemic disease like leukemia, diabetes. A severe osteitis with bone loss could lead to the formation of an oroantral fistula of one or both maxillary sinus.
- *Syphilis*—gummata of the palate may result in a massive oroantral fistula due to destruction of the intervening bone.
- *Inadequate blood clot formation*—inadequate blood clot formation in the alveolus after violation of the maxillary sinus may lead to oroantral communication.
- *Implant denture*—there is destruction of the antral floor in a patient fitted with an implant denture in maxillary region.
- *Malignant granuloma, Wegener's granulomatosis and lymphosarcoma*—when the invasive process spreads to the palate, expansive perforation or ulceration may occur leading to huge fistula.
- *Periodontal problems*—deep periodontal pockets along the side of the upper molar can destroy the bone, which separate the root from the antrum.

Clinical Features

- *Opening*—oroantral opening can be seen immediately after extraction of the tooth.
- *Immediate symptoms*—there is passage of fluid from oral cavity into the nose. Patient also notices inability to blow out cheek and smoke cigarettes. Unilateral epistaxis, due to blood in the maxillary sinus escaping through the nasal ostium is present. There may be escaped of air from the mouth into the nose and an alteration in vocal resonance.

- *Delayed symptoms*—the reason for the delayed response is that, at first instance the oroantral defect was completely occluded by blood clot and it is only when this plug disintegrates as a result of infection than an oroantral communication is firmly established. There may be foul, sweetish, fetid or salty taste in mouth. Facial pain or headache which is of throbbing nature and is exacerbated by movement of the head. There may be painless lump on the gum at the site of extraction socket.
- *Unilateral nasal discharge*—unilateral nasal discharge accompanied by sensation of nasal obstruction or nocturnal coughing resulting from draining of exudate into pharynx.
- *Postnasal drip*—postnasal drip will often lead to an unpleasant taste which is accompanied by nocturnal cough, hoarseness, earache or catarrhal deafness.
- *Acute exacerbation*—inadvertent entry of food particles, chewing gum, fluids, impression materials, dressing and packs may provoke acute exacerbation of the inflammatory disease.
- *Signs of recent fistula*—after forceful extraction, floor of sinus seen with root of the teeth. Sudden disappearance upper molar root while extraction (**Fig. 27-22**). There may be water running out of the nose while rinsing.
- *Signs of established fistula*—a simple dimple on the alveolar ridge may be the only sign. Invasion of antral polyp through fistula resulting in sudden appearance of exophytic mass on the alveolar crest. There is also aspiration of air into mouth through the tooth socket. Tenderness is positive over the maxillary sinus.
- *Sinusitis*—sinusitis occurs with varying intensity depending on diameter of fistula. Patient exhibits swelling and redness covering the sinus and molar eminence as well as pain beneath eye.



Fig. 27-22: Oroantral fistula presented as an opening intraorally (Courtesy Dr Ashok L).

Radiographic Features

- *Break in continuity of maxillary sinus*—radiograph will show break in continuity of maxillary sinus. In some cases there is disalignment of a small portion of the cortical layer of bone.
- *Features of sinusitis*—there are also characteristic features of acute or chronic sinusitis due to ingress of bacteria.
- *Second view should be taken by tilting the head to know location*—evidence of displaced root or tooth and second view of the antrum with the head in a different position may be required to ascertain the exact location of the displaced object.
- *Silver probe examination*—confirmation of presence of fistula can be done by introducing silver probe into the orifice, followed by taking the radiograph.

Diagnosis

- *Clinical diagnosis*—all extracted upper posterior teeth should be examined, not only to ensure they are intact but also for indication of a sinus contamination. If the root is covered with a thin plate of bone or adherent sinus mucosa a communication may be present.
- *Air blow technique*—the patient should be asked to blow air into the pinched nose with the mouth open. This forces the air into the sinus via ostium. If an oroantral opening is present, bubbles appear in the extraction socket.
- *Probing of sinus*—Gentle probing of the socket with a blunt instrument, such as ball ended periodontal probe or small amalgam plugger will confirm the bone defect without perforating an intact lining.
- *Radiological diagnosis*—CT will be able to confirm the diagnosis of oroantral fistula (**Fig. 27-23**).

Management

- *Drug therapy*—antibiotic therapy for 1 week. Most commonly given antibiotics is amoxicillin. Decongestants are given for 1 week.
- *Drainage*—to promote drainage, the oroantral fistula can be enlarged by excising a margin of surrounding mucosa, by cauterizing any granulation tissue and polyps protruding from the sinus with electrocoagulation or simply by cautious application of a chemical agent such as trichloroacetic acid. The patient is provided with a syringe and blunted wide bore needle or a plastic irrigating tube, to wash out the sinus with warm salt water three or four times daily. In some cases, antral lavage and antrostomy is done to help drainage.
- *Prevention from dislodging clot*—if opening is small, great care is to be exercised such as avoidance of use of irrigation, vigorous mouth washing and frequent lusty

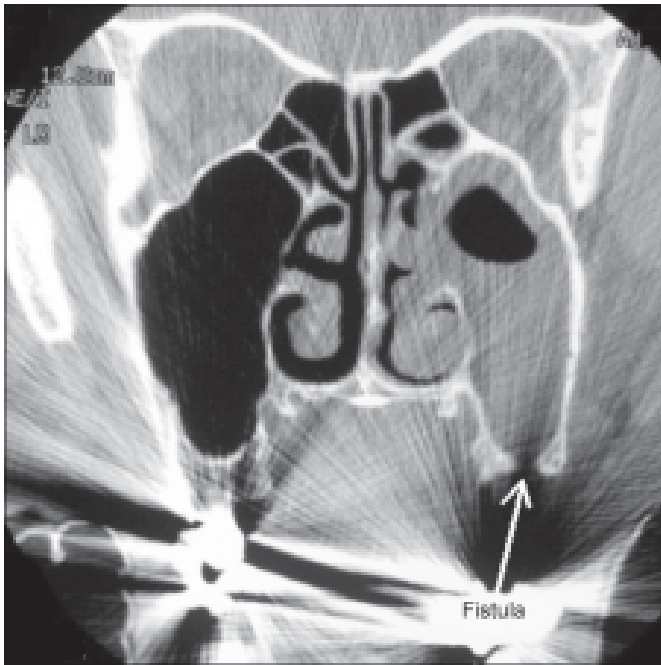


Fig. 27-23: Oroantral fistula seen as an opening on the CT scan (Courtesy Dr Ashok L).

blowing of the nose. In majority of the cases, good clot will form and normal healing will occur.

- *Closure of opening*—if large accidental opening is present, its immediate closure can be achieved in the following ways:
 - *Rising of mucoperiosteum*—mucoperiosteum is raised both buccally and palatally and the height of the alveolar ridge is reduced at the site of opening. Edges of soft tissue are freshened up. A periosteal elevator should be used to raise palatal mucoperiosteum so that approximation of mucosal edges is made possible.
 - *Relaxing incision*—relaxing incision is made over the palate avoiding the palatine artery.
 - *Suturing*—flaps are then sutured without tension.
 - *Shrinkage of nasal mucosa*—nasal drops should be given to shrink nasal mucosa and promote drainage.

Calcification

Antroliths

These are calcified masses found in the maxillary sinus. It is defined as a complete or partial calcific encrustation of an antral foreign body, either endogenous or exogenous which serves as nidus. There is calcification of masses of stagnant mucus in site of previous inflammation, root fragments or bone chips or foreign body.

Etiology

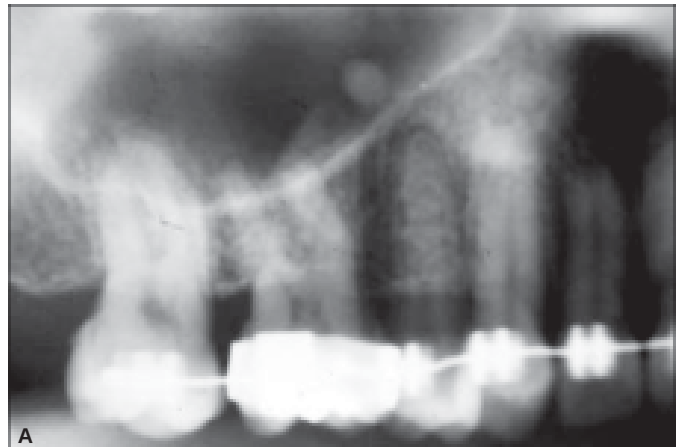
- *Endogenous nidus*—endogenous nidus is usually root tip or may be simply fragment of soft tissue, bone, blood or mucus.
- *Exogenous nidus*—exogenous nidus are consisting of snuff or paper.

Clinical Features

- *Age and sex*—it can occur at any age in either sex.
- *Symptoms*—it is usually asymptomatic. But if they continue to grow the patient may complaint of bloodstained nasal discharge, nasal obstruction or facial pain.

Radiographic Features

- *Radiodensity*—they may have a homogenous or heterogeneous density.
- *Internal structure*—there is alternating layer of radiolucency and radiopacity in the form of lamination.
- *Margins*—outline may be ragged or smooth.
- *Shape*—shape may be round (Figs 27-24A and B), oval or irregular and it is attached to the wall of the sinus.



Figs 27-24A and B: Antroliths seen as round shaped radiopacity in the maxillary sinus (Courtesy Dr Ashok L).

Diagnosis

- *Clinical diagnosis*—it is not possible to make confirmed clinical diagnosis.
- *Radiological diagnosis*—round shaped radiopaque structure in the sinus will give clue to the diagnosis.

Differential Diagnosis

- *Root fragments*—pulp canal is seen and it is moved in different direction if head position is changed.

Management

- Removal of antroliths can be done if it is symptomatic.

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Traumatic Injury of Soft Tissue

Traumatic Erythematous Macule or Hematoma

Traumatic macule is induced by low grade, usually chronic, physical insult.

Clinical Features

- *Site*—common sites are anterior and lateral border of tongue (**Fig. 28-1**), floor of mouth, posterior palate, buccal mucosa and mucosal surface of lip.
- *Symptoms*—mild to considerable pain and it regresses after the cause is removed.
- *Signs*—it may blanch when digital pressure is used.
- *Size*—size of red zone corresponds closely to size of traumatic agent.
- *Margins*—margins may or may not sharply define.



Fig. 28-1: Traumatic hematoma of the tongue occur due to tongue biting.

Diagnosis

- *Clinical diagnosis*—red lesion with history of trauma may give clue to the diagnosis.

- *Laboratory diagnosis*—inflamed lamina propria covered with thin or eroded stratified squamous epithelium that is completely non-keratinized.

Management

- *Removal of irritant*—mechanical irritant is identified and should be removed.

Ecchymosis and Purpuric Macule (early stage)

It is produced by blunt traumatic insult of sufficient force, on the mucosa of skin to cause superficial extravasation of blood (**Fig. 28-2**).



Fig. 28-2: Soft tissue injury due to trauma.

Clinical Features

- *Site*—palate, buccal mucosa, upper lip (**Fig. 28-3**) and floor of mouth are common site.

- **Size**—size varies according to size and force of physical agent inflicting the damage.
- **Margins**—borders are poorly demarcated and blend imperceptibly with surrounding normal tissue.
- **Signs**—it does not blanch on pressure as RBCs are within the tissue rather than in the vessel.



Fig. 28-3: Macular lesion seen on upper lip due to blunt trauma.

Diagnosis

- **Clinical diagnosis**—macular lesion with history of blunt trauma.

Management

- It is not required, if the diagnosis is confirmed.

Cervicofacial Emphysema

It is a swelling occurs due to introduction of gas or air in the interstices of the connective tissue. In orofacial region, forced air may spread to mediastinal and retropharyngeal area.

Etiology

- **Dental procedure**—it follows variety of oral and dental procedures like tooth extraction, blowing of compressed air into the root canal during endodontic treatment or into a periodontal pocket, blowing of air from high speed air rotor machine.
- **Fracture**—it can also follow mid facial fractures.
- **Surgical procedure**—it can occur spontaneously as a result of patient's breathing actions following some type of surgical procedure with the break in tissue permitting air to enter the connective tissue space. In some cases, while surgical operation, compressed air may be forced in small intraoral laceration.

Clinical Features

- **Symptoms**—patient complains of pain and 'bubbling' sensation while palpating the tissue and often difficulty in breathing.
- **Appearance**—it is manifested as unilateral swelling of the tissues of face and/or neck which occurs very rapidly.
- **Crepitus**—it can be felt while auscultating the swelling.
- **Mediastinal involvement**—spread to mediastinum will result in dysphonia, dysphagia or dyspnea.
- **Hamman's crunch**—cardiac auscultation reveals crepitus synchronous with the heart beat in case of mediastinum involvement.
- **Pneumoparotid**—it occurs when air enters parotid duct. It appears as unilateral enlargement of the parotid. Milking of parotid duct produce frothy air filled saliva.

Diagnosis

- **Clinical diagnosis**—history and presence of crepitus on palpation are two important diagnostic indicators for cervicofacial emphysema.

Management

- **Antibiotics therapy**—usually broad spectrum antibiotics are prescribed for dental related cervicofacial emphysema. It usually resolves in 2 to 5 days of time.
- **Management of pneumoparotid**—these are treated with antibiotics, massage, hydration, sialogogues and warm compress.

Factitious Injury

Factitious disorders are characterized by physical or psychological symptoms produced intentionally to assume the sick role. A symptom can be considered intentionally produced if there is direct evidence or if other causes for the symptom have been excluded. A patient presenting with a factitious illness typically has a personality disorder with prominent borderline traits. These patients, when confronted with the possibility that their illness was self-inflicted, typically deny the accusations, become angry. Treatment of this disorder is difficult. Perhaps the best treatment may be recognized that it exists so that unnecessary procedures can be avoided. A factitious patient will rarely seek psychiatric treatment. When our patient was confronted with the possibility that her problem was self-inflicted, she vehemently denied the accusations.

Self-inflicted injury is the most common in subcutaneous emphysema. Therefore, when a patient presents with unexplained recurrent subcutaneous emphysema, one should suspect self-infliction and examine for puncture marks.

There may be circular purpura around the mouth. In some cases patient give injury to area to get relief from the pain (Fig. 28-4). This is more common in rural population.



Fig. 28-4: Factitious injury seen on the neck of patient to get relief from the pain.

Traumatic Injury to Teeth

There are various causes of traumatic injuries to teeth like falls, collisions, sporting activities, domestic violence, automobile accident and assault.

Factors Affecting Trauma to Teeth

- *Energy of impact*—this factor includes both mass and velocity. Low velocity blows cause the greatest damage to the supporting structure whereas the high velocity blows are usually not associated with damage to the supporting structure.
- *Resiliency of impacting object*—if the tooth is struck with cushioned object or if the lip absorb and distribute the impact, the chances of crown fracture are reduced while the risk of luxation and alveolar fracture is increased.
- *Shape of impacting object*—a sharp impact favors clean crown fracture with minimum displacement of the tooth because the energy is spread rapidly over a limited area. On the other hand, a blunt impact increases the area of resistance and force in the crown is transmitted to the apical region causing luxation or root fracture.
- *Direction of the impacting force*—impact can meet the tooth at different angles, most often hitting the tooth facially, i.e. perpendicular to the long axis of the root. Depending upon on the direction of the impacting force the type of fracture properties of enamel and dentin; it was seen that enamel is weakest in region parallel to the enamel rods and that dentin is easily fractured when force is perpendicular to the dentinal tubules.

Classification

Ellis and Davey's classification

- *Class I*—simple fracture of the crown involving enamel.
- *Class II*—extensive fracture of the crown, with considerable amount of dentin involved but no pulp exposure.
- *Class III*—extensive fracture of the crown with considerable amount of dentin involved with pulp exposure.
- *Class IV*—traumatized tooth becomes nonvital (with or without loss of crown structure).
- *Class V*—tooth lost due to trauma.
- *Class VI*—fracture of root with or without loss of crown structure.
- *Class VII*—displacement of the tooth without crown or root fracture.
- *Class VIII*—fracture of crown and mass.
- *Class IX*—fracture of deciduous teeth.

Classification by Andreessen

This classification is based on a system adopted by WHO in its application of the international classification of disease to dentistry and stomatology.

- *Injuries to the hard dental tissues and pulp*
 - *Crown infraction*—an incomplete fracture of the enamel without loss of the tooth substance.
 - *Uncomplicated crown fracture*—it is restricted to enamel or involves enamel and dentin, but does not expose the pulp.
 - *Complicated crown fracture*—a fracture involving enamel and dentin and exposing the pulp.
 - *Complicated crown root fracture*—a fracture involving enamel, dentin and cementum and exposing pulp.
 - *Root fracture*—a fracture involving dentin, cementum and the pulp.
- *Injuries to the periodontal tissue*
 - *Concussion*—an injury to the tooth supporting structures without abnormal loosening or displacement of the tooth, but with marked reaction to percussion.
 - *Subluxation*—an injury to the tooth supporting structure with abnormal loosening but without displacement of tooth.
 - *Intrusive luxation* (central dislocation)—displacement of the tooth into alveolar bone. This injury is accompanied by comminution or fracture of the alveolar socket.
 - *Extrusive luxation* (peripheral dislocation)—partial displacement of the tooth out of its socket.
 - *Lateral luxation*—displacement of the tooth in a direction other than axially. This is accompanied by comminution or fracture of the alveolar crest.
 - *Extra-articulation* (complete avulsion)—complete displacement of the tooth out of its socket.

- *Injuries of the supporting bone*
 - *Comminution of alveolar socket*—crushing and compression of the alveolar socket. This condition was found with intrusive and lateral luxation.
 - *Fracture of the alveolar socket wall*—a fracture contained to the facial or lingual socket walls.
 - *Fracture of the alveolar process*—a fracture of the alveolar process which may or may not involve the alveolar socket.
 - *Fracture of mandible and maxilla*—a fracture involving the base of the mandible or maxilla and often the alveolar process. The fracture may or may not involve the alveolar socket.
- *Injuries to gingiva and oral mucosa*
 - *Laceration of gingiva or oral mucosa*—a shallow or deep wound in the mucosa resulting from a tear and usually produced by a sharp object.
 - *Contusion of gingiva or oral mucosa*—a bruise usually produced by impact from a blunt object and not accompanied by a break of the submucosal hemorrhage.
 - *Abrasion of gingiva or oral mucosa*—superficial wound is produced by rubbing or scrapping of the mucosa leaving a raw bleeding surface.

Classification by Garcia Godoy

- 0—Enamel crack.
- 1—Enamel fracture.
- 2—Enamel, dentin, fracture without pulp exposure.
- 3—Enamel, dentin fracture with pulp exposure.
- 4—Enamel dentin cementum fracture without pulp exposure.
- 5—Enamel dentin cementum fracture with pulp exposure.
- 6—Root fracture.
- 7—Concussion.
- 8—Luxation.
- 9—Lateral displacement.
- 10—Intrusion.
- 11—Extrusion.
- 12—Avulsion.

Concussion

In this case, there is some injury to the supporting structures without abnormal loosening or displacement of teeth and occurs due to crushing injury to the vascular structure at apex and to the periodontal ligament with resulting inflammatory edema.

Clinical Features

- *Symptoms*—traumatized tooth is sore.
- *Signs*—it is sensitive to horizontal and vertical percussion and biting force.

Radiographic Features

- *PDL space widening*—widening of periodontal ligament space in the apical portion.
- *Reduce pulp canal size*—reduction in the size of the pulp canal after a year following the trauma.

Management

- *Conservative*—adjustment of opposing teeth should be done to avoid pain.

Luxation

It is the dislocation or disarticulation of teeth which are abnormally mobile and are displaced. There may be complete or partial necrosis of the pulp.

Types

- *Subluxation*—it denotes abnormal loosening of teeth without frank dislocation.
- *Intrusive luxation*—displacement of teeth into the alveolar bone.
- *Extrusive luxation*—partial displacement of teeth out of the socket.
- *Lateral luxation*—movement other than axial displacement.

Clinical Features

- *Site*—teeth commonly involved are maxillary incisors in both dentitions.
- *Symptoms*—teeth are abnormally mobile. Pain is also present.
- *Signs*—bleeding from the gingival crevice indicating periodontal damage. Tooth is extremely sensitive to percussion and masticatory force. Clinical crown may appear shortened in case of intrusive luxation.
- *Sequelae of luxation*—there may be external root resorption, ankylosis of tooth, obliteration of root canals, pulp necrosis and marginal bone loss.

Radiographic Features

- *Tooth elevation*—elevation of traumatized tooth.
- *PDL space widening*—widening of apical portion of periodontal ligament space.
- *Pulp chamber obliteration*—in intrusion, partial or complete obliteration of pulp chamber may occur.

Management

- *Subluxation*—if several teeth are traumatized or subluxated, a splint may be placed to stabilize the involved teeth during the healing phase. Follow-up care

is mandatory for these injuries because of the greater potential for pulpal necrosis. A splint can be removed within 7-10 days.

- *Extrusive luxation*—the clinician first repositions the luxated tooth into its alveolar socket. If a clot has formed apical to the displaced tooth, the tooth may be more difficult to reposition and more manipulation and pressure may require. It requires splint to be used to stabilize the tooth during the healing process for 2 to 3 weeks. The tooth should be examined clinically and radiographically at monthly interval. If the periapical lesion appears or a fistula is present, endodontic therapy becomes a major consideration.
- *Lateral luxation*—the repositioning of laterally luxated tooth requires a more forceful degree of reduction because of the type or displacement that has occurred. Manipulation with the thumb and index finger can often reduce the injured tooth. The laterally luxated tooth should be repositioned, first by forcing the displaced apex out of its locked position within the labial bone, allowing the clinician to place axial pressure in an apical direction and to manipulate the tooth into its natural position. Splinting is required routinely after reduction of lateral luxation injuries. Maintain the splint for minimum of 14 days and remove it when no abnormal mobility is associated with the tooth.
- *Intrusive luxation*—the best policy is to wait and watch for the tooth to re-erupt on its own accord. Surgical and orthodontic extrusion should be done. Root canal therapy must be anticipated in almost all intrusive luxation injuries.

Avulsion

It is complete displacement of tooth from its alveolus.

Clinical Features

- *Age*—encountered in a relatively younger age
- *Site*—maxillary central incisors are commonly affected. Teeth may be missing from the arch (**Fig. 28-5A**).
- *Associated signs*—fracture of the alveolar wall and lip injuries are frequently seen.

Radiographic Features

- *Socket without tooth*—presence of dental socket without correspondent tooth. Tooth may be located in the adjacent soft tissue.
- *Noticeable lamina dura*—if recent wound is seen, lamina dura will be noticeable.
- *Socket sclerosis*—socket sclerosis occurs.



Figs 28-5A and B: Missing tooth from the arch in Figure A and sample tooth is kept in saline solution in Figure B (Courtesy Dr Shetty).

Management

- *Management at site of injury*—replant immediately, if possible. If contaminated, rinse with water before implanting. When immediate implantation is not possible, place the tooth in the transport Medias like milk, saline (**Fig. 28-5B**), saliva and if none of the above is present, use water.
- *Replantation of tooth*—it refers to the insertion of a vital or non-vital tooth into the same alveolar socket from which it was removed or otherwise lost. If extra-oral dry time is less than 2 hours, replant immediately and of extraoral dry time is greater than 2 hours then soak in a topical fluoride solution for 5-20 minutes, rinse with saline and replant. If tooth has been in physiological storage media, then replant immediately.
- *Management of the root surface*—keep the tooth moist at all times and do not handle the root surface. If the root surface is contaminated, rinse with saline and if persistent debris remains on the root surface then use

cotton pellet to remove the remaining debris and/or gently brush off the debris with a wet sponge.

- *Management of socket*—gently aspirate without entering the socket. If a clot is present, use light irrigation with saline. Do not make surgical flap unless bony fragment prevents replantation.
- *Root canal treatment*—mature tooth with complete root formation must have root canals filled before implantation or else pulp necrosis will result. If root canals are not filled, there is gradual obliteration of the pulp chamber and canals by calcification.
- *Splinting*—splinting can be done by using a variety of appliance including stainless steel wires, acrylic splints, orthodontic banding and arch wires with wire ligatures and even surgical cements with gauze. In some cases root resorption begins within a week or to a few months or as long as 10 years.
- *Transplantation of teeth*—it is the replacement of teeth which is damaged beyond repair by caries or by means of other teeth. Usually, mandibular first molar is replaced by developing third molar. Transplanted teeth have to be esthetically acceptable with no periapical or lateral lesions, sharing adequately in the maintenance or physiologic maxillomandibular and muscular relations. Homologous transplants of preserved frozen teeth have also been proposed to simplify the process.

Bruxism

The word *bruxism* is taken from the Greek word *brychein*: gnashing of teeth. Although the term *bruxism* is not generally known to lay people, it is shorter and more convenient than *teeth clenching or grinding*. *Bruxism* can perhaps be best defined as the involuntary, unconscious and excessive grinding, tapping, or clenching of teeth. When it occurs during sleep, it may be best called as *sleep or nocturnal bruxism*. A few people, on the other hand, brux while they are awake, in which case the condition may be called *wakeful or diurnal bruxism*.

All forms of bruxism entail forceful contact between the biting surfaces of the upper and lower teeth. It is also called as '*night grinding*' or '*bruxomania*'. It is a habitual grinding of the teeth, either during sleep or as an unconscious habit during waking hours. It is the term used both for clenching habit during which pressure is exerted on the teeth and periodontium by the actual grinding or clamping of the teeth and, also to repeated tapping of the teeth.

Etiology

- *Local*—mild occlusal disturbances, unconscious attempt by the patient to establish a greater number of teeth in contact or to counteract a local irritating situation.
- *Systemic factors*—gastrointestinal disturbances, sub-clinical nutritional deficiency and allergy or endocrine disturbances have been reported to be the causative factors.
- *Psychological factors*—emotional tension in which patient is unable to express his emotion due to fear, rage, rejection and it becomes hidden in subconscious and later expressed by variety of way like by grinding the teeth.
- *Occupational*—like in cases of watchmaker, persons who chew gum, tobacco or objects such as toothpicks or pencils.

Clinical Features

- *Symptoms*—in grinding and tapping, this is contact which involves movements of the lower jaw and unpleasant sounds which can often awaken housemates. Clenching (or clamping) on the other hand, involves inaudible, sustained, forceful teeth contact unaccompanied by mandibular movements. Long-term bruxers sometimes experience jaw tenderness, jaw pain, fatigue of facial muscles, headaches, neckaches, earaches and hearing loss.
- *Teeth*—chronic bruxism may lead to sensitive, worn-out, decayed, fractured, loose or missing teeth. Grinding or clenching breaks down the enamel; sometimes in long-term bruxers, the teeth height is reduced to stumps (Fig. 28-6). Instead of a white enamel cover, one often sees the more yellowish and softer dentin. The posterior teeth of some chronic bruxers often lose their cusps and natural contours, appearing instead flat, as if they had been worked over with a file or sandpaper. When anterior teeth are affected, their biting surfaces are damaged.



Fig. 28-6: Attrition seen lower anterior region in chronic bruxers.

- *Dental caries formation*—the absence of enamel makes it easier for the bacteria to penetrate the softer part of the teeth and produce cavities. With time, the condition may lead to bridges, crowns, root canals, implants, partial dentures and even complete dentures. As long as bruxism continues, the situation keeps getting worse. Thus, by 40 or 50 years of age, most patients have worn their teeth to the degree that extensive tooth restorations have to be performed.
- *Appearance*—as the teeth wear out, they become shorter. As a result, when the mouth is closed, the upper and lower jaws are nearer than they used to be and hence the nose and chin. The skin now may bag below the eyes and curl around the lips, causing the lips to seemingly disappear. The chin recedes and the person looks comparatively old.
- *Facial muscles*—bruxism involves excessive muscle use leading to a build-up or enlargement (hypertrophy) of facial muscles, especially those of the jaws where the masseter muscle (the muscle that raises the lower jaw and enables closing the jaws) is located. In long-term bruxers, this build-up may lead to a characteristic square-jaw appearance. Some patients resort to removing part of the masseter muscle by surgery or injections of toxic materials to reduce muscle size and thus partially regain their former, more aesthetically pleasing look.
- *Salivary glands*—another example of this spiral involves the occasional inflammation and blockage of some salivary glands. In this case, the masseter muscle becomes disproportionately overdeveloped and blocks the opening of the nearby parotid glands. They, thus interfere with the flow of saliva into the mouth, causing the saliva to accumulate in the glands. This in turn may lead to periodical swelling, pain, inflammation and abnormal dryness of mouth.
- *TMJ*—bruxism may also damage the temporomandibular joints. First few signs of temporomandibular joint disorders are TMJ discomfort or pain, soreness of jaws and muscles, clicking or popping sounds when opening the jaws or while chewing and difficulties in opening the mouth fully.
- *Malocclusion*—malocclusion or bad bite is more common among bruxers than in the general population. Bruxism may often involve more pressure on one side of the mouth than on the other, thereby causing malocclusion. As the teeth wear out (**Fig. 28-7**), the distance between the upper and lower jaw decreases and overclosure may develop.
- *Effect on periodontium*—There may be loss of integrity of the periodontal structures resulting in loosening or drifting of the teeth and even gingival recession occurs.



Fig. 28-7: Severe destruction tooth seen in bruxism patient.

Management

- *Psychotherapy*—the belief that bruxism is traceable to stress and other emotional and psychological factors give rise to a variety of psychotherapeutic approaches. For instance, listening to progressive relaxation or autosuggestion tapes just before going to sleep may foster calmness and self-confidence.
- *Wakeful EMG Feedback*—another psychological approach to stress reduction resorts to instrumentation. During bruxing, the relevant muscles are active and this increased activity or tension can in turn be measured with an electromyograph (EMG: electro—electric; myo—muscle; graph—record). During treatment sessions at home or the laboratory, the patient sits or reclines comfortably. One or more pairs of recording electrodes are then attached to the surface of the skin in close contact to appropriate muscles (e.g. masseter muscles). These electrodes transmit information about the level of muscle activity to a computer monitor. The patient is instructed to consciously lower that level below a threshold line (also visible on the screen). Gradually, by becoming alert to the presence of muscle tension, patients may develop techniques for reducing that tension and hence, bruxism.
- *Exercise*—Quinn suggested isokinetic and stretching exercises of the mandible. Such exercises may or may not help alleviate bruxism, but perhaps may be used to complement other approaches. However, but it seems unlikely that they could ever be used as the sole therapeutic approach. Evidences that this approach is effective and non-existent.
- *Drugs*—both, the stress and brain malfunction etiological theories give at times, rise to the use of anti-anxiety agents, muscle relaxant and other drugs. Most authorities, however, feels that at best, drugs in use now are of limited value in the treatment of great majority of

chronic bruxers and that they often involve moreover untoward side effects. Evidence that this approach is effective and are non-existent.

- *Equilibration therapy*—some people believe that bruxism is traceable to malocclusion (bad bite). They therefore suggest eliminating this cause through orthodontic adjustment.
- *Splints*—by far, the most common treatment regimen for bruxism relies on the time-honored procedure of splints like nightguards, biteguards, occlusal splints, biteplates, removable appliances or interocclusal orthopedic appliances and use of manufactured customized appliances. Removable splints are worn at night to guide the movement so that periodontal damage is minimal.

Fracture of Teeth (Fig. 28-8)

Dental Crown Fracture

Anterior teeth are commonly involved. It may be caused by fall, accident and blows from foreign bodies.

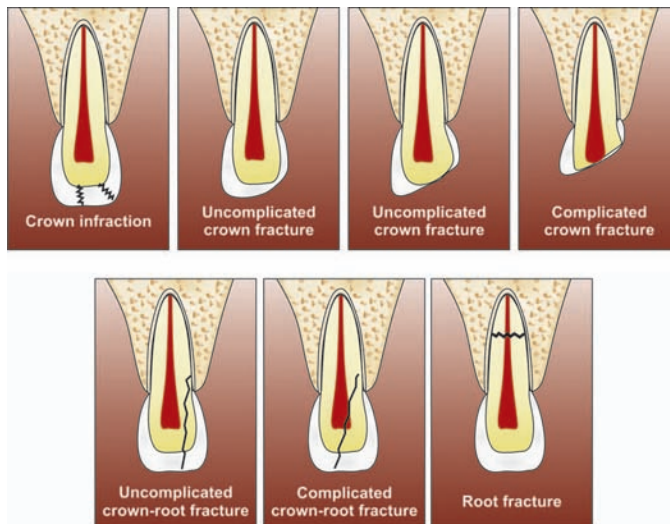


Fig. 28-8: Injuries to hard dental tissues and pulp.

Types

- *Enamel infraction or crack*—fracture that involves only enamel without loss of enamel substance.
- *Uncomplicated fracture*—fracture involving enamel or enamel and dentin with loss of tooth substance without pulpal involvement.
- *Complicated fracture*—fracture that extends through enamel, dentin and pulp with loss of tooth substance.

Clinical Features

- *Cracks*—it can be seen in indirect light (directing the beam along the long axis of tooth).



Figs 28-9A and B: Uncomplicated fracture of crown of teeth (Courtesy Dr Shetty).

- *Uncomplicated fracture*—it does not involve pulp and is usually found on mesial or distal corner of maxillary central incisors (Figs 28-9A and B). Dentin involvement is identified by contrast of color between it and peripheral layer of enamel. Exposed dentin is very sensitive to chemical, thermal and mechanical stimulation.
- *Complicated fractures*—there is bleeding from exposed pulp or at least drop of blood oozes from the pinpoint exposure. Exposed pulp will be sensitive to most forms of stimulation (Figs 28-10A and B).

Radiographic Features

- *Appearance*—it will show size and position of pulp chamber, location and extent of exposure and stage of root development.

Management

- *Crown infraction*—no treatment (Fig. 28-11).
- *Uncomplicated fracture*—small defect is corrected by grinding. Oblique fracture has worse prognosis than



Figs 28-10A and B: Complicated crown fracture of upper central incisor (Courtesy Dr Shetty).

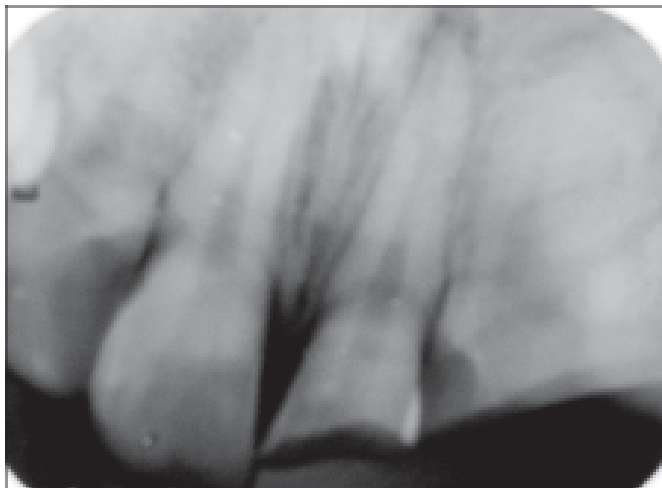


Fig. 28-11: Crown fracture involving the enamel and dentin.

horizontal fracture. Provide a temporary glass-ionomer cement bandage or permanent restoration using a bonding agent and composite resin. If very close to pulp, consider $\text{Ca}(\text{OH})_2$ base. If an intact fragment exists, a bonding procedure may be carried out.

- **Complicated fracture**—pulp capping, pulpotomy and pulpectomy can be done. In immature tooth: Perform pulp capping or partial pulpotomy with calcium hydroxide and bacteria tight coronal seal. In mature tooth: Same as with immature tooth or pulpectomy.

Dental Root Fracture

Clinical Features

- **Sites**—it is common with maxillary central incisors.
- **Coronal fragment**—coronal fragment is displaced lingually and is slightly extruded. If fracture line is close to the apex then tooth will be more stable (**Fig. 28-12A**).
- **Signs**—if only movement of crown is detected, root fracture is likely to be considered.
- **Symptoms**—temporary loss of sensitivity. It returns to normal within 6 months.



Fig. 28-12A: Root showing fracture at the apex of root (Courtesy Dr Soni).

Radiographic Features

- **Appearance**—if the X ray beam is projected parallel with the plane of the root, sharp radiolucent line between the fragments can be seen (**Fig. 28-12B**). If the X-ray beam is not parallel, it will look as poorly defined gray shadow.
- **Fracture line**—fracture line may be transverse or oblique.
- **Obliteration of pulp chamber**—calcification and obliteration of pulp chamber can be seen in due course of time.

Differential Diagnosis

- **Soft tissue image**—it extends beyond tooth margin.

Management

- **Fracture in middle or apical third of root**—teeth mainly restored to their proper position and securely immobilized.
- **Fracture at coronal/cervical area**—extraction is indicated.

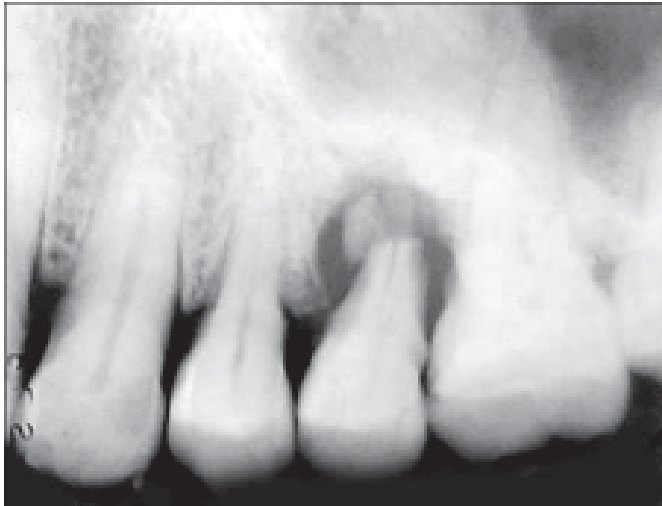


Fig. 28-12B: Fracture root in apical region.

Crown/Root Fracture

Such fracture is likely to be intra- and extra-alveolar. They are result of direct trauma.

Clinical Features

- *Site*—they have labial margin in the gingival third and course obliquely to exist below the gingival attachment on the lingual surface. It frequently involves pulp (Fig. 28-13).
- *Symptoms*—there is pain during mastication.
- *Signs*—tooth is sensitive to occlusal force.



Fig. 28-13: Crown/root fracture of upper central incisor due to severe attrition of tooth.

Radiographic Features

- *Appearance*—most crown/root fractures are perpendicular to the direction of the beam.

Management

- *Removal of coronal fragment*—this can be done and after this, dowel crown should be prepared.
- *Extraction*—if 3 to 4 mm of root is remaining, extraction is indicated.
- *Uncomplication fracture*—uncomplicated fracture is to be restored.

Vertical Root fracture

It is also called as 'Cracked tooth syndrome'. It runs length-wise from crown towards apex of tooth.

Causes

- *Iatrogenic*—they are iatrogenic in origin, i.e. after insertion of pin or retention screws into vital and non-vital teeth.
- *Endodontically treated tooth*—it also occurs in endodontically treated tooth as there is weakening of tooth in such cases.
- *Traumatic occlusion*—it may be caused by traumatic occlusion.

Clinical Features

- *Site*—it is usually seen in posterior teeth in adults, especially in mandibular molars.
- *Symptoms*—dull pain of long duration which may vary from non-existent to mild.
- *Signs*—it may have periodontal lesions resembling chronic lesion.
- *History*—history of repeated failure of endodontic treatment.

Radiographic Features

- *Appearance*—if central X-ray lies in the plane of the fracture, it may be visible as a radiolucent line.
- *PDL thickening*—after bacterial invasion, radiograph will show thickening of periodontal ligament space or a diffuse radiolucent lesion.

Management

- *Extraction*—extraction of single rooted teeth.
- *Hemisection*—hemisection for multi-rooted tooth followed by extraction of involved half and the remaining half is restored with endodontic therapy.

Perforation of the Root

The root canal is sometimes penetrated during operative procedures and injury may extend to cause a perforation of the root. The usual site of it is at the apex and the side of the root, but the floor of the pulp chamber and even the side of

the crown; near the neck of the tooth may be perforated (Fig. 28-14).

Radiographically, it is visible if some smooth metal broach or piece of wire or some opaque substance is inserted in the abnormal passage to make it visible. In some cases, the shadow of root canal can be seen approaching the side of the root and this can be the evidence of a perforation. Rarefying osteitis at the side of a tooth is always suggestive of perforation, particularly if there has been root canal therapy.

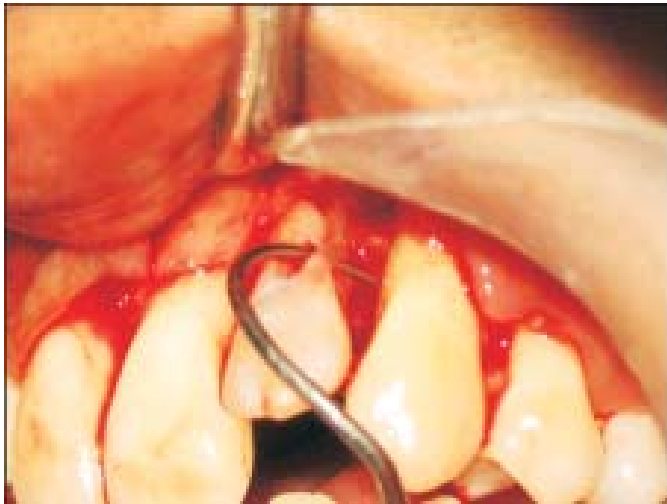


Fig. 28-14: Root perforation seen in the central incisor (Courtesy Dr Soni).

Tooth Ankylosis

Ankylosis between tooth and bone is an uncommon phenomenon in the deciduous dentition and even more rare in permanent teeth.

Causes

- *Partial root resorption*—it occurs when partial root resorption is followed by repair; with either cementum or bone that unite the tooth root with alveolar bone.
- *Occlusal trauma*—ankylosis can occur after traumatic injury particularly due to occlusal trauma.
- *Following root canal therapy*—it may follow root canal therapy, if the apical periodontal ligament is irritated or seriously damaged.

Clinical Features

- *Symptoms*—it seldom manifests any clinical symptoms unless there is concomitant pulp infection.
- *Dull muffled sound*—if the extensive area of the root surface is involved, the tooth gives a dull muffled sound on percussion rather than the normal sharp sound.
- *Difficulty in extraction*—there is considerable difficulty while doing extraction of the teeth, sometimes necessitating surgical removal.

Radiological Features

- *Loss of PDL space*—it may be visible on the radiograph as loss of the normal thin radiolucent line representing the periodontal ligament.
- *Sclerosis*—there may be mild sclerosis and apparent blending of the bone with the tooth root.

Management

- It has good prognosis and as such no treatment is required.

Traumatic Injury to Facial Bone

When patient is subjected to injury, there is chance that jaws may be fractured. Even in some cases, minor trauma may result in bone damage.

Radiographic Features of Fracture

There are mainly following features seen in fracture of jaws:

- *Fracture line*—dark line which results from the passage of rays through the line of cleavage has been termed as the fracture line. Fracture line is very dark, when the rays pass freely between the fragments and less dark when the rays pass through small part of the fracture line.
- *Displacement of the fragment*—in it, instead of normal continuity of the surface of bone, there is dis-alignment of the two fragments. In mandible, action of temporalis muscle pulls the posterior portion of the mandible upward is the common cause of displacement.
- *Deformity*—it is alteration in shape of bone as a result of injury in the cortex. The abrupt angulations of bone are indication of deformity.
- *Radiopaque line*—a fracture that has become impacted does not have any space between the fragments and the line of cleavage is obscured by superimposition of bony trabeculae at the site of impaction. In this case, there is slight increase in density at the fracture line due to double number of trabeculae.
- *Double fracture line*—many fractures pass slight obliquely across the bone. When the line of cleavage of inner and outer cortices does not superimpose, it gives an impression that there are two fractures.
- *Relationship of teeth to fracture line*—a fracture may enter the apical end of the tooth socket and disappear. If the line of fracture extends along a lateral aspect of the tooth, the tooth may obscure it. Fracture through the angle of the jaw frequently appears to extend from the apices of the 3rd molar but it has actually started at the alveolar margins of bone. An unerupted tooth may sometimes be present in the fracture line, which might play as predisposing factor in fracture and its presence may

affect the treatment. With fracture, a tooth may be forced from its follicle into the adjacent bone, soft tissue or between the fragments; causing its separation.

Indication for Removal of a Tooth from the Fracture Line

- Longitudinal fracture involving the root.
- Dislocation or subluxation of the tooth from the socket.
- Presence of periapical infection and infected fracture line.
- Acute Pericoronitis, advanced periodontitis.

Dentoalveolar Fractures

These are defined as those in which avulsion, subluxation or fracture of teeth occur in relation with fracture of the alveolus. Anterior alveolar fractures are most common. Labial plate is more prone to fracture than palatal plate. Fracture line is most often horizontal.

Clinical Features

- *Site*—alveolar process of maxilla is more commonly fractured than that of mandible. The most common sites are the upper incisor and cuspid regions and region of tuberosity.
- *Lip*—there is full thickness wound of the lower lip or ragged laceration on its inner aspect caused by impaction against the lower anterior teeth. There may be local bruising and portions of teeth or foreign bodies may get lodged in soft tissue of lip.
- *Gingiva*—there may be laceration of gingiva and deformity of alveolus.
- *Teeth*—there may be fracture of crown and root portion of teeth. Teeth in fractured fragment will have recognizable dual sound when percussed.
- *Occlusion*—marked malocclusion along with displacement and mobility.
- *Alveolar fracture*—a complete alveolar fragment may be displaced into the soft tissue of the floor of mouth and can be covered by mucosa. There may be gross comminution of alveolus occurs.
- *Maxillary tuberosity fracture*—the detached bone may include the floor of maxillary sinus (**Fig. 28-15**); which is indicated by discharge from the nose on the involved side. Ecchymosis of buccal vestibule can also occur.
- *Cracked pot noise*—impacted alveolar fracture may be virtually immobile. Sometimes crepitations can be detected on palpation and a 'cracked pot' noise is detected when the teeth within the alveolar fracture are percussed.

Radiological Features

- Radiologically, fracture line is seen above the apices of root of teeth.



Fig. 28-15: Fracture of maxillary tuberosity showing detachment.



Fig. 28-16: Dentoalveolar fracture showing line distal to central incisor.

- Damage to roots of the adjacent teeth is common with fracture of alveolus (**Fig. 28-16**). Fracture of alveolar process is well delineated. Posterior palatal fracture is best demonstrated on occlusal radiograph.

Management

- *Fracture of maxillary tuberosity*—if the tuberosity is completely detached from the periosteum, it should be carefully dissected out and the resulting soft tissue defect are sutured to prevent any residual opening into the maxillary sinus.
- *Extensive fracture of alveolus with teeth attached*—splinting of teeth and anchoring to teeth elsewhere in the upper jaw.

Mandibular Fractures

Fracture of mandible occurs more frequently than any other fracture of the facial skeleton. It is more common than middle third fracture. The most common facial fractures are mandible (61%), followed by maxilla (46%), the zygoma

(27%) and the nasal bones (19%). Most common cause of mandibular fracture is road traffic accidents. Other causes are assault, falls, industrial trauma and sport injury.

Classification

- *According to type of fracture:*
 - *Simple*—it includes closed linear fracture of the condyle, coronoid, ramus and edentulous body of the mandible. Greenstick fracture occurring in children is also included in it.
 - *Compound*—fracture of tooth bearing portions of the mandible are nearly always compound into the mouth via the periodontal membrane and some severe injuries are compound through the overlying skin.
 - *Comminuted*—direct violence to mandible from penetrating sharp objects and missiles are usually compound and may be further complicated by bone and soft tissue loss.
 - *Pathological*—when fractures result from minimum trauma to mandible which is already weakened by pathological conditions like osteomyelitis, neoplasm and generalized skeletal diseases.
- *According to site of fracture:*
 - Dentoalveolar
 - Condyle
 - Coronoid
 - Ramus
 - Angle
 - Body (molar and premolar)
 - Parasymphysis
 - Symphysis
- *According to cause of fracture:*
 - *Direct violence*—fracture occurs at the site of trauma.
 - *Indirect violence*—fracture occurs away from the site of trauma.
 - *Excessive muscular contraction*—occasionally, fractures of coronoid process occur because of sudden reflex contracture of temporalis muscle.
- *According to treatment plan*
 - *Unilateral*—it is more frequently caused by direct violence.
 - *Bilateral*—it occurs due to combination of direct and indirect violence.
 - *Multiple*—it is also associated with direct and indirect violence. Most common fracture of this type is Guardsman fracture when soldier faints on parade at midpoint of chin causing fracture of symphysis and both the condyle.
 - *Comminuted*—it is usually seen in war missile injury (Fig. 28-17).
- Fry and colleagues classified, in descriptive manner, the different directions of the fracture:

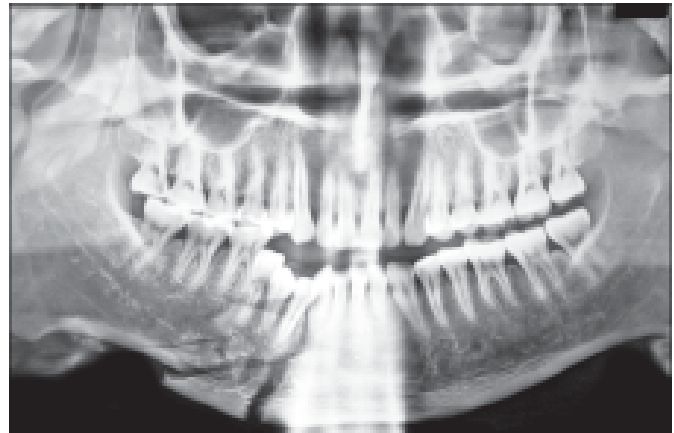


Fig. 28-17: Comminuted fracture of body of mandible.

- *Horizontal favorable*: if the direction of the fracture is such that it resists the action of the elevator muscles in pulling the ramus segment upwards when viewed from side, or horizontal plane called as horizontally favorable fracture.
- *Horizontally unfavorable*: when the direction of fracture line is reversed and posterior fragment is elevated by the muscle pull, the fracture called as horizontally unfavorable (Fig. 28-18).
- *Vertically favorable*: when viewed from above or in the vertical plane, the buccolingual direction of the fracture line is such that it prevents the displacement of the segment lingually by resisting the pull of the medial pterygoid muscle called as vertically favorable.
- *Vertically unfavorable*: when the opposite is the case, in which the segment can be pulled lingually because of the unopposed action of the medial pterygoid, the fracture is termed as vertically unfavorable.



Fig. 28-18: Horizontally unfavorable fracture of the angle of mandible and parasymphyseal fracture involving the teeth in fracture line.

Clinical Features of Mandibular Fracture

- *Changes in the occlusion*—any change in the occlusion is highly suggestive of the mandibular fracture. A change in the occlusion can result from fractured teeth, a fractured alveolar process, a fractured mandible at any location, and trauma to the temporomandibular joint and muscles of mastication. Posttraumatic premature posterior dental contact or anterior open bite may result from bilateral fractures of the mandibular condyle or angle as well as from maxillary fractures with inferior displacement of the posterior maxilla. Posterior open bites may occur with the fractures of the anterior alveolar process or parasymphiseal fractures. Unilateral open bite may occur owing to ipsilateral angle and parasymphiseal fracture (**Fig. 28-19**). Posterior cross bite can result from midline symphyseal and condylar fractures with splaying of the posterior mandibular segments.



Fig. 28-19: Step deformity seen in the fracture of mandible in the symphysis region.

- *Anesthesia, paresthesia, or dysesthesia of the lower lip*—Numbness in the distribution of the inferior alveolar nerve after trauma is almost pathognomonic of a fracture distal to the mandibular foramen.
- *Abnormal mandibular movement*—most patients presenting with a fractured mandible have limited opening and trismus owing to guarding of muscles of mastication. Certain mandibular fractures result in predictable abnormal mandibular movements like deviation on opening towards the side of a mandibular condylar fracture (**Fig. 28-20**). Because lateral pterygoid muscle function on the unaffected side is not counteracted on the opposite side by the nonfunctioning of lateral pterygoid muscle, deviation results. Inability to close the jaws can be the result of fractures of the alveolar process, angle, ramus, and symphysis, causing



Fig. 28-20: Fracture mandible showing deviation on opening.

premature dental contact. Lateral mandibular movements may be inhibited by bilateral condylar fractures and fracture of ramus with bone displacement.

- *Change in facial contour and mandibular arch form*—facial asymmetry should alert clinician to the possibility of mandibular fracture. A retruded chin can be caused by bilateral parasymphiseal fracture. The appearance of an elongated face may be the result of bilateral subcondylar, angle, or body fracture, allowing the anterior mandible to be displaced downward. If there is a deviation from the normal U-shaped curve of the mandible, fracture should be suspected.
- *Lacerations, hematoma and ecchymosis*—trauma significant enough to cause loss of skin or mucosal continuity or subcutaneous-submucosal bleeding certainly can result from trauma to the underlying mandible. Lacerations should be carefully inspected before closure. The direction and type of fracture may be visualized directly through the laceration. In rare cases of animal bites there may be the loss of soft as well as hard tissue. The diagnostic sign of ecchymosis or hematoma in the floor of the mouth indicates mandibular body or symphyseal fracture.

Radiographic Features

- *Technique required*—following types of radiographs are helpful in the diagnosis of mandibular fractures, i.e. panoramic (**Figs 28-21 and 28-23**) radiograph, lateral oblique radiograph, posteroanterior radiograph, occlusal view, periapical view, reverse Towne's view, CT scan.
- *Margin of fracture*—the margins of the fractures usually appear as sharply defined radiolucent lines of separation that are confined to the structure of the mandible. Occasionally the margins of the fracture overlap each other, resulting in the area of increased radiopacity at the fractured site.



Fig. 28-21: Fracture at angle of mandible on left side.



Fig. 28-23: Fracture at symphysis and angle of mandible.



Fig. 28-22: Fracture at body of the mandible.



Fig. 28-24: PA view showing multiple fractures of the mandible.

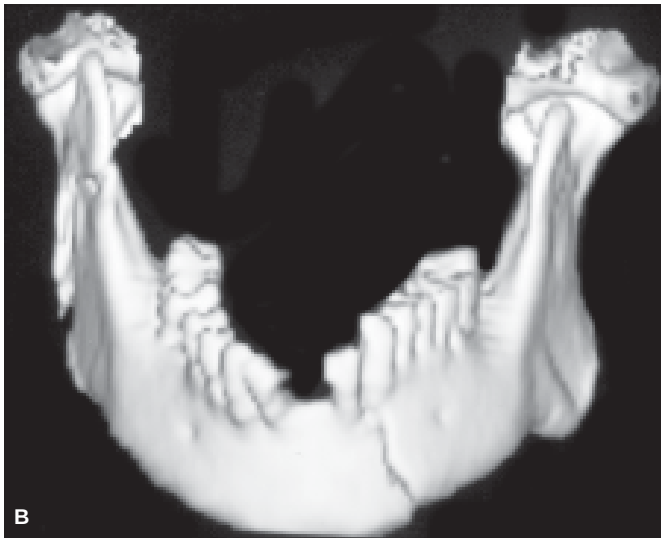
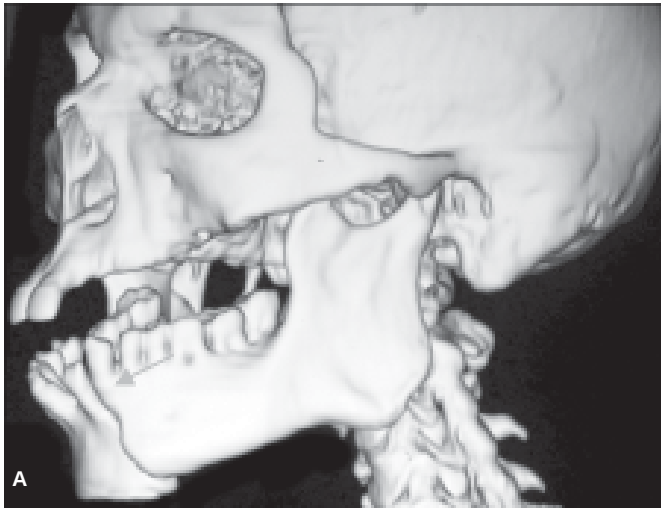
- *Step*—displacement of the fracture results in the cortical discontinuity or “step.”
- *Double fracture line*—an oblique fracture that involves both cortical plates may show two fracture lines if the fracture lines in both buccal and lingual plates are not superimposed. In this case, a right angle view such as occlusal view and the fact that the two fracture lines are meeting at same point on the inferior border of the mandible helps in correction of diagnosis.
- *Lateral oblique view*—the lateral oblique view of the mandible can be helpful in the diagnosis of ramus, angle and posterior body fractures (Fig. 28-22).
- *PA view*—the posteroanterior view demonstrates any medial or lateral displacement of the fractures of ramus, angle, body or symphysis (Fig. 28-24).
- *Occlusal view*—the mandibular occlusal view demonstrates displacement in the lateral or medial direction of the body fractures and also shows the anterior or posterior displacement of the symphyseal fracture.
- *Reverse Towne’s view*—the reverse Towne’s view is ideal for showing medial displacement of the condyle and

condylar neck fracture. Periapical dental film shows the most detail and can be used for the nondisplaced linear fractures of the body as well as alveolar process and dental trauma.

- *CT scan*—the CT scan is ideal for the condylar fractures that are difficult to visualize. The greater expenses and radiation exposure limit its use in cases that cannot be diagnosed with conventional radiograph (Figs 28-25A to 28-26).

Differential Diagnosis

- *Soft tissue shadow of space between tongue and soft palate*—in lateral radiograph of normal mandible, it is possible to see the gray shadow of tongue which has curved upper and posterior borders. Immediately behind lies the soft palate, which presents a shape somewhat similar to an inverted triangle. In many radiographs, there is a dark line situated between these two shadows which is



Figs 28-25A and B: 3D CT image of mandibular fracture at symphysis region (Courtesy Dr Amit Parate, Lecturer, GDCH, Nagpur, India)

produced by presence of air between the tongue and soft palate. This thin dark line is carried down over the shadow of the mandible in the region of the angle and sometimes gives resemblance to a fracture. Careful study shows that the dark line is continued on the bone.

- *The shadow of hyoid bone*—in lateral radiograph of jaws, the shadow of neck intervenes over the some part of the bone and where it ends anteriorly, there is difference in the radiographic density which is quite marked in some cases. There is greater likelihood of making error when there is thin dark line present which infact represents the subcutaneous fat on anterior aspect of the neck. In either case, there is evidence of shadow continuing on the bone and no fracture can produce such an effect.
- *The intervertebral disc space*—it presents as dark shadow and can be mistaken for fracture. In this case, intraoral occlusal film can be placed in the mouth in occlusal

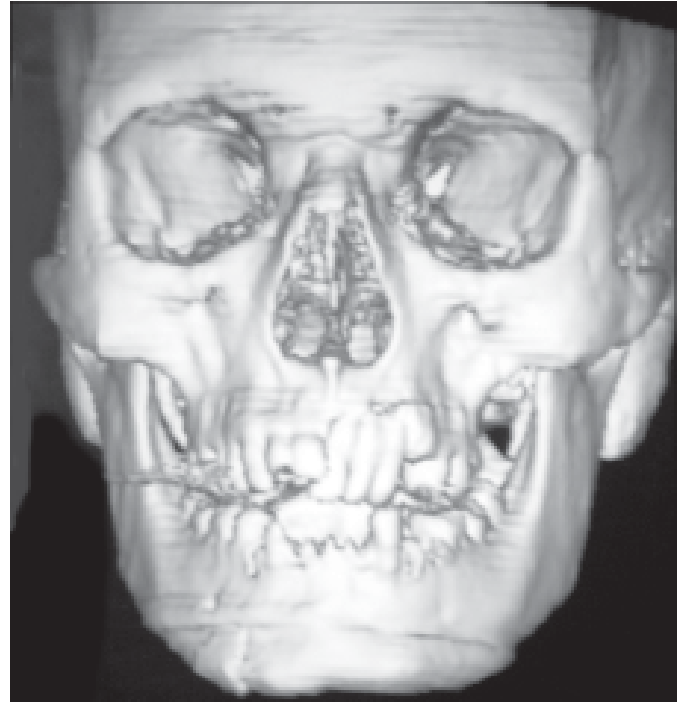


Fig. 28-26: 3D CT of fracture of mandible (Courtesy Dr Meshram).

plane and X-rays are directed in an appropriate manner, to produce excellent radiographs of this region.

Management

Preliminary management

- *Examination*—the first aid required consists of careful examination of mouth and removal of all fragments of teeth, broken filling and dentures.
- *Airway*—if there is danger of falling tongue back, then dorsum of the tongue should be sutured.
- *Hemorrhage*—obvious bleeding point such as facial vessels should be secured with artery forceps and temporary dressing should be applied.
- *Soft tissue laceration*—soft tissue wound should be sutured within 24 hours of injury.
- *Antibiotics*—benzyl penicillin should be administered IM injection or 1 mega unit every 6 hours for first 2 to 3 days and oral penicillin should be continued for further one week. In recent, oral metronidazole 400-800 mg BD is given to all patients with mandibular fracture.

Planned management

- *Reduction*—reduction of fracture means the restoration of functional alignment of bone fragments.
- *Immobilization of fractured bone*—the fracture site must be immobilized to allow bone healing to occur. Immobilization can be done with intermaxillary fixation and bone plating.

- *Minimally displaced fracture*—close reduction and intermaxillary wiring (**Fig. 28-27**).
- *Severely displaced fracture*—open reduction and intermaxillary fixation.



Fig. 28-27: Fracture with interdental wiring is seen.

Postoperative Care

- *Sitting position*—patient with fracture of mandible finds it more comfortable if they are in sitting position with chin forward, provided that there is no contraindication to this posture such as fracture of vertebra.
- *Drugs*—morphine and its derivative are contraindicated in patients with maxillofacial injuries as these drugs depress the respiratory center and cough reflex. Prophylactic antibiotics should be administered to control the infection. Maintenance of good oral hygiene is essential for prevention of infection.

Coronoid Fracture

It is rare. It usually results from the reflex contracture of the powerful anterior fibers of temporalis muscle. During operation on large cyst of ramus, coronoid may get fracture (**Fig. 28-28**).



Fig. 28-28: Fracture of coronoid is also seen in association with fracture of condyle and mandibular body.

If the tip of the coronoid process is ditched, the fragment is pulled upwards towards the infratemporal fossa by the temporalis muscle. There may be tenderness over the anterior part of the ramus and hematoma. Painful limitation of movement, especially on protrusion of mandible may be found.

Middle Third Fractures

Classification

- *Central middle third fracture*
 - *Le Fort's type-I*—bilateral detachment of the alveolar process and palate or the low level subzygomatic fracture of Guerin.
 - *Le Fort's type-II*—pyramidal subzygomatic fracture of the maxilla.
 - *Le Fort's type-III*—high level suprazygomatic fracture of central and lateral part of face.
- *Fracture of zygomatic complex*
 - Zygoma depressed with fracture at several sites
 - Fracture of zygomatic arch
- *Fracture of naso-ethmoidal complex*
- *Fracture of orbit*
 - Fracture of orbital rim
 - Orbital blow out fracture

Midfacial fracture

It may involve the frontal, nasal, lacrimal, zygoma, ethmoidal and sphenoid bones. They are classified by Rene Le Fort.

Le Fort I

It is also known as low level fracture/Horizontal fracture of Maxilla/Guerin fracture/Floating fracture.

- Horizontal fracture above the level of Nasal floor (**Fig. 28-29**).

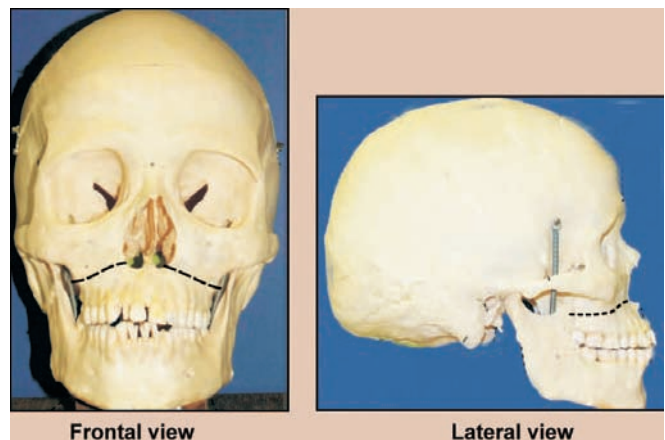


Fig. 28-29: Diagrammatic representation of Le Fort I fracture.

- Fracture line extends backward from the lateral margin of the anterior nasal aperture below the zygomatic buttress to cross the lower 3rd of the pterygoid lamina.
- Fracture also passes along the lateral wall of the nose and the lower 3rd of nasal septum to join the lateral fracture behind the tuberosity.

Le Fort II

It is also called as 'Pyramidal or Subzygomatic fracture'. There is a pyramidal appearance of fracture in PA skull view (Fig. 28-30).

- This fracture runs from the thin middle area of the Nasal Bones down to either side crossing the frontal processes of maxilla into the medial wall of each orbit.
- Within each orbit, the fracture line crosses the lacrimal bone behind the lacrimal sac before turning forward to cross the infraorbital margin slightly medial to or through the infraorbital foramen.
- The fracture line now extends downwards and backwards across the lateral wall of the antrum below zygomatico-maxillary suture and divides the pterygoid laminae about halfway up. Separation of block from the base of skull is completed via the nasal septum and may involve the floor of the anterior cranial fossa.

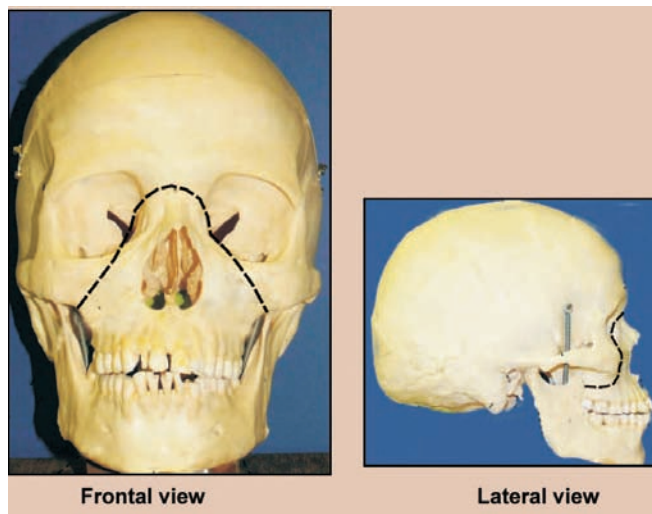


Fig. 28-30: Le Fort II fracture—A diagrammatic representation.

Le Fort III

It is also called as 'High transverse or Suprazygomatic fracture'. It completely separates the middle third of the facial skeleton from the cranium (Fig. 28-31).

- The fracture line runs from near the frontonasal suture transversely, parallel with the base of the skull and involves the full depth of the ethmoid bone, including the cribriform plate.
- Within the orbit, the fracture line passes below the optic foramen into the posterior limit of the inferior orbital fissure.

- From the base of inferior orbital fissure, the fracture line extends into two directions:
 - Backwards across the pterygomaxillary fissure to fracture line the root of the pterygoid laminae.
 - Laterally across the lateral wall of the orbit separating the zygomatic bone from the frontal bone.

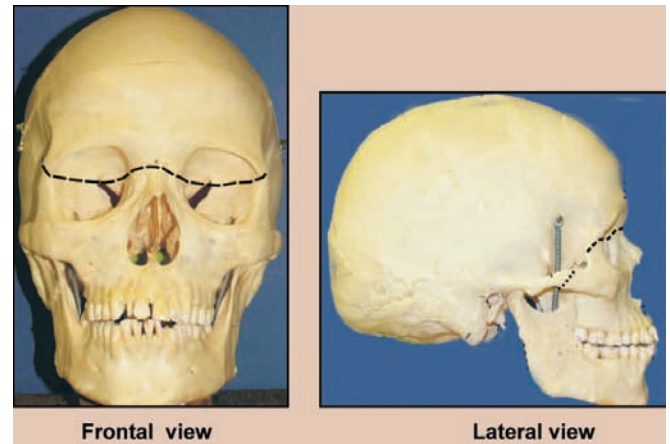


Fig. 28-31: Le Fort III fracture—A diagrammatic representation.

Clinical Features

Le Fort I

- **Location**—it may be unilateral or bilateral.
- **Symptoms**—in recent injury, there may be slight swelling of upper lip. Some fractures are so mobile that the patient has to keep the mouth slightly open to accommodate the increased vertical dimension of bite. Paresthesia presents over the distribution of infra-orbital nerve and pain over nose and face.
- **Signs**—ecchymosis is present in the buccal sulcus beneath the zygomatic arch. Occlusion is disturbed and variable amount of mobility may be found in the tooth bearing segment of maxilla. If the fracture line is at high level, the fragment will include the pterygoid muscle attachment. It will pull the fragment posteriorly and depress its posterior margin. Due to this, posterior maxillary teeth will force the mandible open resulting in open bite, retruded chin and long face.
- **Cracked pot sound**—in impacted type of fracture, there may be damage to cusps of individual teeth caused by impaction of the mandibular teeth against them. Percussion of upper teeth results in a distinctive 'Cracked pot' sound.
- **Flattening of face**—in some cases, there is flattening of middle of the face and epistaxis.
- **Palpation**—manipulation will reveal mobile maxilla and crepitation.

Le Fort II

- **Moon face appearance**—there is massive edema that causes marked swelling of the middle third of face giving rise to 'moon face' appearance.

- **Hemorrhage**
 - *Bilateral circumorbital ecchymosis*—ecchymosis about the eye which develops within minutes of injury (**Fig. 28-32**).
 - *Subconjunctival ecchymosis*—the conjunctivae over the inner quadrant of eye are blood shot and if the zygomatic bone is involved, this subconjunctival ecchymosis will also be over the outer quadrant.
 - *Periorbital hematoma*—it causes swollen conjunctiva to bulge out from between the eyelids.



Fig. 28-32: Circumorbital edema seen in maxillary fracture.

- **Eye**—there may be variation in the size of pupil which may occur due to peripheral damage to the oculomotor nerve. There may be diplopia and ocular movements may be limited.
- **Nose**—there may be lengthening of the nose. There may be bleeding from the nose.
- **Face**—there is 'dish-face' deformity of the face with occasional lengthening of the face.
- **Nerve damage**—as the fracture line passes across the inferior orbital rim, there may be injury to the infraorbital nerve resulting in anesthesia or paresthesia of cheek.
- **Step deformity**—there is step deformity at infraorbital margin. By applying the pressure between the bridge of the nose and the palate, pyramid of bone can be moved.
- **Cerebrospinal fluid rhinorrhea**—it is sometimes appreciated by salty taste in the mouth.
- **Intraoral finding**—retropositioning of the maxilla, so that anterior teeth do not meet and there is gagging on the posterior teeth. There is mobility of the upper jaw. Occasional hematoma of palate, which will cause patient considerable discomfort. Cracked pot sound on tapping the upper teeth.

Le Fort III

- **Location**—superficially, it appears similar to Le Fort II but injury is much more severe. It is very unusual to find Le Fort III fracture occurring in isolation.

- **Edema**—it is more extensive and massive.
- **Signs**—there is tenderness and often separation at the frontozygomatic sutures. The facial skeleton will be tilted to the side opposite to the direction of the fracturing force. There may be mobility of whole of the facial skeleton as a single block.
- **Nose**—nose often blocked with blood clot. There may be cerebrospinal fluid (rhinorrhea). There may be lengthening of nose.
- **Cerebrospinal fluid rhinorrhea**—it will be profused as compared to Le Fort II.
- **Hooding of eye**—there is lowering of the ocular level, due to the fracture line passing above the Whitnall's tubercle, removing the support given to the eye by Lockwood's suspensory ligament. As one or both eye drops, the upper lid follows the globe down, producing unilateral or bilateral 'hooding' of the eyes.
- **Hemorrhage**—bleeding into periorbital tissues.
- **Face**—there is separation of both frontozygomatic sutures which produces lengthening of face.
- **Step deformity**—flattening and step deformity at infraorbital margin.
- **Intraoral findings**—the entire occlusal plane may drop producing anterior open bite. There is gagging of occlusion in the molar area.

Radiographic Features

- **Le Fort I**—it is identified on PA, lateral skull and Water's projection (**Fig. 28-33**). Both the maxillary sinuses are cloudy and may show air filled level. Lateral view shows slight posterior displacement of fragment.



Fig. 28-33: Water's view showing Le Fort I fracture with fracture of zygomatic bone.

- *Le Fort II*—it will reveal the fracture of the nasal bone and both, frontal processes of maxilla and infra-orbital rims on both sides or separation of zygomatic sutures on both sides. Deformities and discontinuation of lateral walls of both sides of maxillary sinus (**Fig. 28-34**). There is thickening of the lining mucosa and clouding of maxillary sinus.
- *Le Fort III*—hazy appearance due to soft tissue swelling. Separation of sutures, i.e. of nasofrontal process, maxillofrontal, zygomatico-frontal and zygomatico-temporal. Nasal bone, frontal process of maxilla, both orbital floors and pterygoid plate show radiolucent lines and discontinuity. Ethmoidal and sphenoid sinuses are cloudy.



Fig. 28-34: CT scan showing the fracture at anterior and posterior wall of maxillary sinus.

Management

- *LeFort I*—Low level—intermaxillary fixation. High level—intermaxillary fixation and cranio-maxillary fixation.
- *LeFort II*—reduction followed by intermaxillary fixation. Open reduction and inter-osseous wiring of infraorbital rims. Antibiotics should be given to the patient.
- *LeFort III*—control hemorrhage and maintain airway. Surgery should be delayed until edema subsides. External immobilization should be done.

Zygomaticomaxillary Complex Fracture

The most common causes of ZMC fractures include interpersonal altercations, falls, motor vehicle accidents, and sports injuries. 25% of patients have other associated facial fractures (**Fig. 28-35**).

Classification

- *Class I*—Fracture of zygomatic bone with minimum or no displacement.



Fig. 28-35: Extensive soft tissue injury associated with fracture of zygomaticomaxillary complex.

- *Class II*—Fracture of zygomatic bone with displacement.
 - *Type A*—Rotation around a vertical axis
 - Inversion of the orbital ring
 - Eversion of the orbital ring
 - *Type B*—Rotation around a longitudinal axis
 - Medial displacement of frontal process
 - Lateral displacement of frontal process
- *Class III*—Enbloc displacement of the bone
 - Medial displacement
 - Inferior displacement
 - Lateral displacement
- *Class IV*—Comminution of the zygomatic bone
- *Class V*—Fracture of zygomatic arch alone

Clinical Features

- *Sign*—flattening of the cheek, swelling of the cheek, periorbital hematoma, subconjunctival hemorrhage (**Fig. 28-36**) and limitations of ocular movements, diplopia, ecchymosis and tenderness intraorally over zygomatic buttress, enophthalmos.
- *Symptoms*—lowering of pupil level, tenderness over orbital rim and frontozygomatic suture, epistaxis. Limitations of mandibular movements. Anesthesia of cheek, temple, upper teeth and gingiva. Possible gagging of back teeth on injured side.
- *Others*—step deformity of infraorbital margin. Separation of frontozygomatic suture.

Radiological Features

- Traditional facial radiographs like submentovertex view offers excellent resolution of the zygomatic arches (**Fig. 28-37**).



Fig. 28-36: Subconjunctival hemorrhage seen in Zygomatico-maxillary complex fracture.

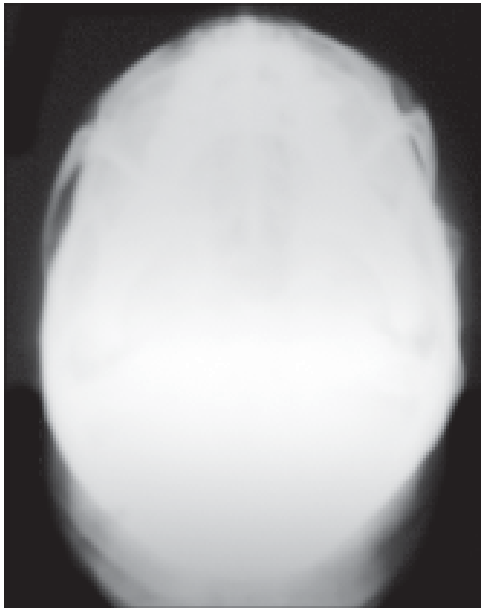


Fig. 28-37: Zygomatic arch fracture.

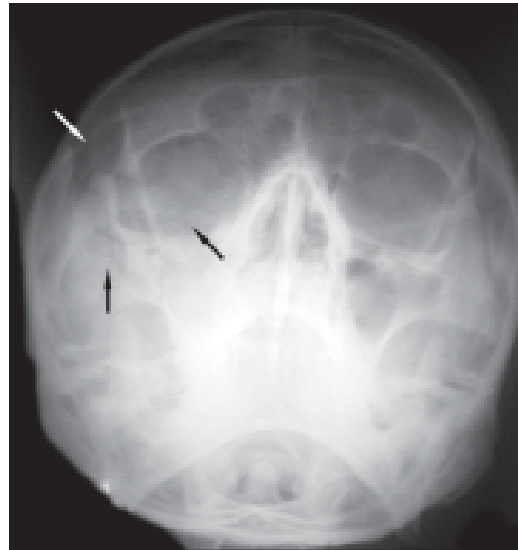


Fig. 28-38: Water's view showing the fractured zygoma.



Fig. 28-39: CT scan, axial section showing the fracture of the anterior wall of maxillary sinus.

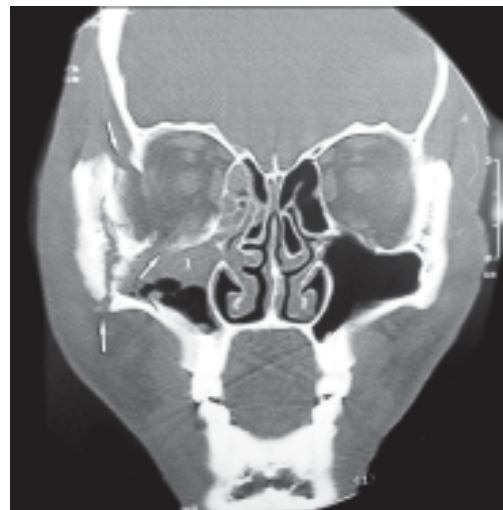


Fig. 28-40: CT scan coronal section showing the lateral displacement of fractured zygomatic bone fragment.

- Other views like Towne's, Water's and the AP views also offer significant information. The occipitontal or Water's view provide good visualization of whole of the zygoma and the maxillary sinus (**Fig. 28-38**).
- CT scans are considered as the standard criteria for the diagnosis of ZMC fractures. The CT scan helps the surgeon to make a more accurate preoperative diagnosis and helps to formulate a treatment plan (**Figs 28-39 and 28-40**).

Management

- **Reduction**—the treatment objective of the management of the zygomaticomaxillary complex fracture is to restore the premorbid malar and orbital configuration. The fracture of the arch and zygoma may be reduced through intraoral or extraoral approach. Three-point reduction is necessary for proper anatomic alignment of the pyramid—shape malar fragment in all planes and adequate fixation must be followed to keep the alignment stable. When there is minimum displacement of the zygomatic bone or arch, no cosmetic deformity or impairment of the eye movement no treatment may be required (**Fig. 28-41**).

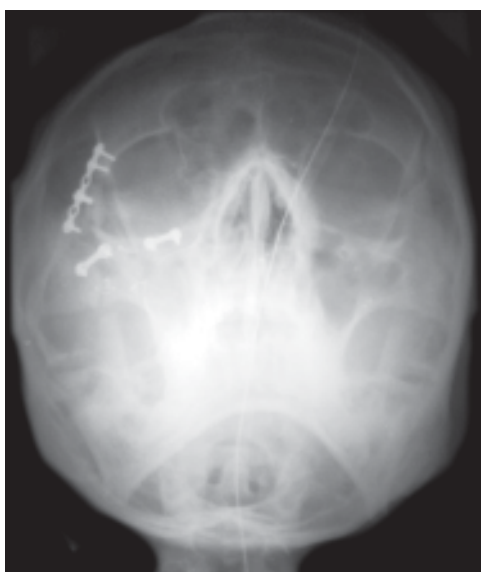


Fig. 28-41: Postoperative radiograph of the patient of zygomatic fracture.

- **Open reduction**—the indications for the open reduction of the zygomaticomaxillary complex fracture are orbital deformities causing ocular disturbance, diplopia, facial asymmetry, multifragmentation and trismus.

Radiological Differential Diagnosis of Fractures of Maxilla

- **Infraorbital foramina**—it often leads to difficulty in diagnosing maxillary fracture when there is a history of blow. The foramen is situated sufficiently below the margin of orbit to form continuous line of bone between the orbit and margin of the foramen. But sometimes, the shadow of the actual canal is thrown over inferior orbital margin and this dark line suggests that a fracture is present. But, continuity of margin can be traced.
- **Suture line between malar bone and maxilla**—in PA view, there is often a thin dark line on the outer wall of antrum, vertical in direction and situated where the inferior margin of the malar bones joins the facial bone. It

represents suture line between the malar bone and maxilla. But in it, there is a thin white line on each side of the dark one and these represent the cortex of the suture.

- **Normal vascular channels**—in intraoral periapical (IOPA) radiograph of the posterior teeth, thin dark lines are often seen in the antral shadows. These represent the normal vascular channels and must not be mistaken for fracture. They all show thin white borders which the fractures do not.
- **Lip line**—in the upper cuspid region, lip line can lead to confusion for fracture line. The lip is retracted at the time that the radiographs are made, adding an extra thickness of tissue in the region where the lip is superimposed.

Greenstick Fracture

It occurs in young people. Rare in maxilla and mandible. Radiologically, there is sharp angulation present at the site of fracture. There may be one or more linear dark streaks running some distance along the length of the bones from each side of the angulation. Small spicules of bone may be seen standing away from the surface, which had been stripped away at the time of fracture.

Nasoethmoidal Injuries

An area behind which lies the interorbital space, which is situated between the medial walls of orbits (**Fig. 28-42**). Fractures in this region are invariably comminuted.

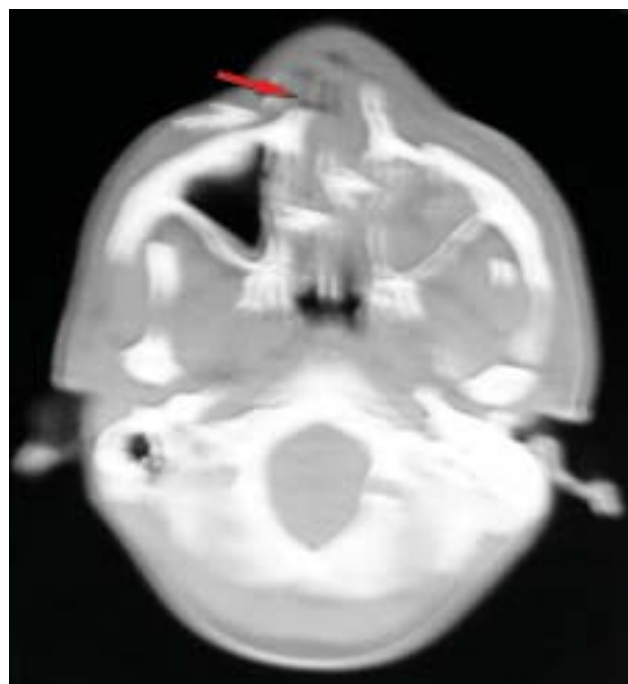


Fig. 28-42: CT scan showing fracture of nasoethmoidal fracture.

Classification

Isolated nasoethmoidal injury

- *Bilateral*—central midface injury resulting from direct blow over nasal bridge. Base of nose is driven backwards into interorbital space and nasal tip becomes upturned. Deep crease at base of nose and skin at base of nose frequently lacerated. CSF rhinorrhea should always be suspected.
- *Unilateral*—unilateral nasal deformity. Side of nose is depressed and there is underlying fracture of ethmoid bone.

Combined nasoethmoidal injury plus midface fractures

- *Bilateral*—nasoethmoid complex fracture combined with Le Fort II and Le Fort III fractures. Causes traumatic telecanthus and elongation of midface.
- *Unilateral*—nasoethmoid complex injury plus severe comminution of orbit and zygomatic complex. Unilateral displacement of medial canthal ligament resulting in displacement of eye downwards and laterally.

Clinical Features

It includes frontal bone depression, nasal deformity, traumatic telecanthus, CSF fluid rhinorrhea, diplopia, and hemorrhage from anterior or posterior branches of ethmoidal artery.

Management

- *Closed reduction*—the use of transnasal wires and compression plates is often unsatisfactory.
- *Open reduction*—realignment of bony fragments under direct vision especially at early stages gives better results.
- *Repair of bony skeleton*—Nasal bridge re-attached to frontal bone. All bone fragments must be preserved, aligned and either directly wired or plated with micro plates.

Complications of Healing

In Fracture

- *Non-union*—it results when callus of osteogenic tissue, over each of the two fragments, fails to meet and fuse or when endosteal formation of bone is inadequate. It can occur due to infection of the fracture site, inadequate immobilization and unsatisfactory apposition of bone. Radiograph shows rounding off and sclerosis of the bone ends, a condition known as 'eburnation'. It is common in elderly patients.
- *Fibrous union*—it occurs as a result of lack of immobilization of the damaged bone. The fractured ends united

by fibrous tissue but there is failure of ossification. They may produce pseudoarthrosis.

- *Lack of calcification*—it occurs in dietary deficiency or mineral imbalance.

In Extraction

Dry Socket

It is also called as 'Alveolitis sicca dolorosa', 'Alveolagia or Postoperative osteitis'. It is basically focal osteomyelitis in which blood clot has disintegrated or is lost. It is called as dry socket as after the clot is lost, the socket gives dry appearance because of exposed bone.

Etiology

- *Difficult extraction*—difficult or traumatic extraction, usually removal of impacted mandibular 3rd molars.
- *Dislodgement of clot*—dislodgment and disintegration of the clot and subsequent infection of the bone.

Clinical Features

- *Duration of occurrence*—it is the most common complication in healing of extraction wound. It arises within the first few days after extraction.
- *Symptoms*—it is extremely painful and odor is foul, but with no suppuration.
- *Signs*—exposed bone is necrotic and sequestration of fragments is common.
- *Healing*—healing of such infected wounds is extremely slow.

Prevention

- *Experiment by placing sulfanilamide*—sulfathiazole cones placed in fresh tooth socket of dog can prevent the occurrence of dry socket but it will cause retarded blood clot formation and even cause some breakdown of clot and remarkable delay in epithelialization.
- *Oxidase cellulose*—inserted for hemostatic purpose, produces retardation of healing similar to that of combined sulfonamide.
- *Sulfathiazole* in 60% glycerin base reduces the frequency of occurrence of dry socket.
- *Aureomycin*—causes significant reduction in decomposition of blood clot. There is decreased incidence of postoperative pain and swelling after one week.
- *Tetracycline hydrochloride*—if tablet is placed in extraction sockets, it helps in reduction of dry socket to 0.78%.
- *Trypsin*—digests necrotic tissue and debris and restrains bacterial growth.
- *Antiseptic mouth rinses*—use phenolated antiseptic mouth-rinses prior to extraction.
- *Little trauma*—one should try to cause a little trauma to tissue as possible. So that it helps in normal healing.

- *Myospherulosis*—it is the complication of healing of an extraction wound or soft tissue wound into which there is placement of antibiotic ointment with petroleum base. It results in formation of clear spaces within the area of healing and the presence of altered erythrocytes which assume the appearance of solitary or clusters of spherules that have been mistaken for large microorganism.

Management

- *Packing material*—insertion of packing material containing an obtundent.
- *Zinc oxide and eugenol pack*—iodoform gauze with zinc oxide and eugenol.

In Wound

Fibrous Healing of a Wound

It is an uncommon complication following a difficult, complicated or surgical extraction of tooth. It occurs most frequently when tooth extraction is accompanied by loss of both, lingual and labial cortical plates of bone with accompanied loss of periosteum.

It is usually asymptomatic and discovered only on radiological examination. Radiologically, it appears as a well circumscribed radiolucent area at the site of extraction wound.

Excision of the lesion for the purpose of establishing the diagnosis and to produce normal healing with subsequent bony repair.

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Definition

The deposition of calcium salts, primarily calcium phosphate usually occurs in the skeleton. When it occurs in unorganized fashion in soft tissue, it is referred to as *Heterotopic calcification*.

Classification of Calcification

- *Dystrophic calcification*
 - General dystrophic calcification of the oral regions
 - Calcified lymph nodes
 - Dystrophic calcification in the tonsil
 - Cysticercosis
 - Calcified carotid artery
- *Idiopathic calcification*
 - Sialoliths
 - Phleboliths
- *Metastatic calcification*
 - Ossification of the stylohyoid ligament
 - Osteoma cutis
 - Myositis ossificans.

Dystrophic Calcification

General Dystrophic Calcification of the Oral Region

When calcium salt precipitate into primary site of chronic inflammatory dead and dying tissue, it is called as dystrophic calcification. It is associated with high local concentration of phosphatase, as in normal bone calcification and with anoxic conditions within the devitalized tissue.

Clinical Features

- *Location*—it is most frequent form of pathologic calcification and found in a wide variety of tissues like areas of tuberculosis, necrosis, and blood vessels in arteriosclerosis, scars and areas of fatty degeneration. In oral cavity, area of dystrophic degeneration is found in the gingiva, tongue or cheek.
- *Symptoms*—it is asymptomatic.
- *Signs*—overlying tissue is enlarged and ulcerated and solid mass of calcium salt palpable.

Radiographic Features

- *Site*—the common sites are long standing and chronically inflamed cyst.
- *Appearance*—appear as fine grains of radiopacities which are sparse and diffuse (**Fig. 29-1**).



Fig. 29-1: Dystrophic calcification seen superimposed on ramus of mandible (Courtesy Dr Parate).

- **Size**—may be single or multiple and rarely exceed 0.5 cm in diameter.
- **Margins**—outline is irregular or indistinct.

Calcified Lymph Nodes

These are types of dystrophic calcification that occurs in lymph nodes following chronic infections. Lymph node calcification is uncommon, and usually results from a previous granulomatous disease.

Causes

- *Tuberculosis lymphadenitis*—tuberculous lymphadenitis is probably a disease in which calcified lymph nodes occur.
- *Other diseases*—actinomycosis, histoplasmosis, cat-scratch fever and other chronic inflammatory diseases may also present calcified lymph nodes.

Clinical Features

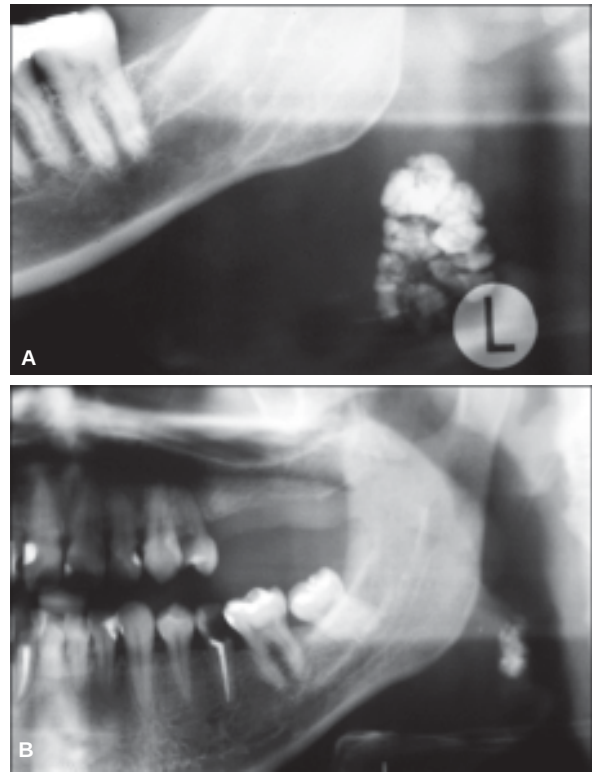
- **Site**—nodes involved are submandibular or cervical chain.
- **Symptoms**—it is usually asymptomatic.
- **Number**—it may be single or multiple or sometimes chain of nodes.
- **Size and shape**—they are hard, round or oblong masses. Outline is well contoured and well defined.
- **Fixity to underlying tissue**—they are mobile during palpation.

Radiographic Features

- **Site**—it is most commonly seen behind or below the angle of the mandible (**Fig. 29-2A**). In some cases, it is seen in more inferior location when cervical lymph nodes are involved (**Fig. 29-2B**). In rare cases, calcified node is found posterior to the ramus.
- **Radiodensity**—it is opaque and well defined. In some cases radiodensity is variable showing both opaque and radiolucent appearance.
- **Appearance**—it may have laminated appearance. Radiopacity often exhibits a patchy pattern with a reticular arrangement of radiolucent lines or gaps.
- **Mass of coral appearance**—sometimes, irregular heterogeneous opaque masses are seen which appear to resemble a mass of coral.
- **Margins**—well defined and usually irregular, occasionally having a lobulated appearance similar to outer shape of cauliflower.

Diagnosis

- **Clinical diagnosis**—it is not possible to make clinical diagnosis.



Figs 29-2A and B: Calcified lymph nodes, cervical lymph nodes that is cauliflower shaped and chainlike (Courtesy Dr Enzo Rovigatti).

- **Radiological diagnosis**—mass of coral appearance and in some cases, laminated appearance.

Differential Diagnosis

- *Sialoliths*—it is painful and has a smooth outline whereas calcified lymph nodes are usually irregular and sometimes lobulated.
- *Phleboliths*—they are smaller and have concentric radiopaque and radiolucent rings.

Management

- **Surgical removal**—large calcified lymph nodes should be surgically removed.

Tonsillolithiasis

It is dystrophic calcification in the tonsil. It is also called as 'Tonsillar calculi', 'Tonsil concretions', and 'Tonsilloliths'. The mechanism is similar to that of calcified lymph nodes. It occurs due to recurrent inflammation of tonsil.

Clinical Features

- **Age and sex distribution**—it is common in older age groups between 20 to 68 years of age. Women are more commonly affected.
- **Location**—it can be unilateral or bilateral.

- *Symptoms*—small calcification produce no symptoms but larger calcification can produce pain, swelling and dysphagia.
- *Recurrent tonsillitis*—it is also common feature of tonsillar calculi.
- *Palpation*—there is hard, yellow submucosal mass of the affected tonsil.

Radiographic Features

- *Site*—it is seen in middle portion of the mandibular ramus in the region where the image of the dorsal surface of tongue crosses the ramus.
- *Appearance*—it can produce 'speckled' appearances of multiple radiopaque superimposed bilaterally on the images of the mandibular rami in a panoramic radiograph.
- *Size*—it may reach the size of 0.5 cm to 14.5 cm.
- *Margins*—multiple small ill defined radiopacities are seen.
- *Internal structure*—it appears more radiopaque than the cancellous bone and same as cortical bone (Fig. 29-3).

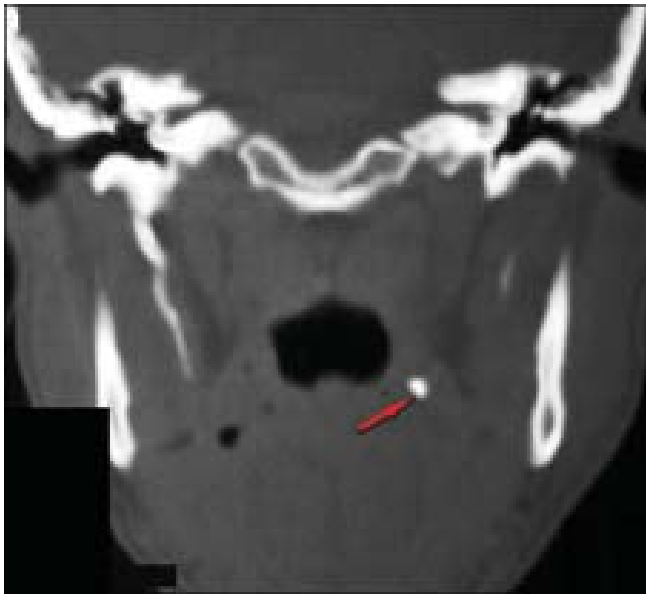


Fig. 29-3: Tonsillitis is present (red arrow) on CT scan (Courtesy RS Kamikawa).

Diagnosis

- *Clinical diagnosis*—recurrent tonsillitis is common features
- *Radiological diagnosis*—speckled appearance with multiple radiopaque image on ramus area.

Differential Diagnosis

- *Radiopaque lesion in mandibular ramus*—in it right angle view such as PA skull or open Towne's view may show that calcification lies to the medial aspect of the ramus.

Management

- *Enucleation*—larger calcifications with associated symptoms are removed surgically.

Cysticercosis

When eggs or gravid proglottids from *Taenia sodium* (pork tapeworm) are ingested by human, their covering is digested in stomach and larval form (*Cysticercus cellulosae*) of the parasite is hatched. Larvae penetrate the mucosa, enter the blood vessels and lymphatics and are distributed in the tissue all over the body. After the larva die, the larval spaces are replaced with fibrous connective tissue, which may become calcified.

Clinical Features

- *Mild cases*—they are completely asymptomatic.
- *Severe cases*—there is mild to severe GIT upset with epigastric pain, severe nausea and vomiting.
- *Nervous system*—convulsion, irritability and loss of consciousness.
- *Size*—palpable firm mass upto 1 cm in diameter.
- *Oral finding*—multiple small nodules may be felt in the region of masseter and suprahyoid muscles and in buccal mucosa or lip.

Radiographic Features

- *Site*—the muscle of mastication and facial expression, the suprahyoid muscle and the postcervical musculature.
- *Margins*—the margins are well defined.
- *Shape*—the shape is elongated, elliptical or ovoid.
- *Internal structure*—it is homogenous and radiopaque.

Diagnosis

- *Clinical diagnosis*—Gastrointestinal problems are present. Small nodules seen in masseter region will give clue to the diagnosis.
- *Radiological diagnosis*—homogenous radiopaque mass is present.

Differential Diagnosis

- *Salivary stone*—it is not multiple as compared to cysticerci.

Management

- *Medical management*—medical management by the physician.

Calcified Carotid Artery

It is also called as 'atheroma'. Arterial wall may calcify in all forms of arteriosclerosis with deposition of calcium salts within the medial coat of the vessels.

Causes

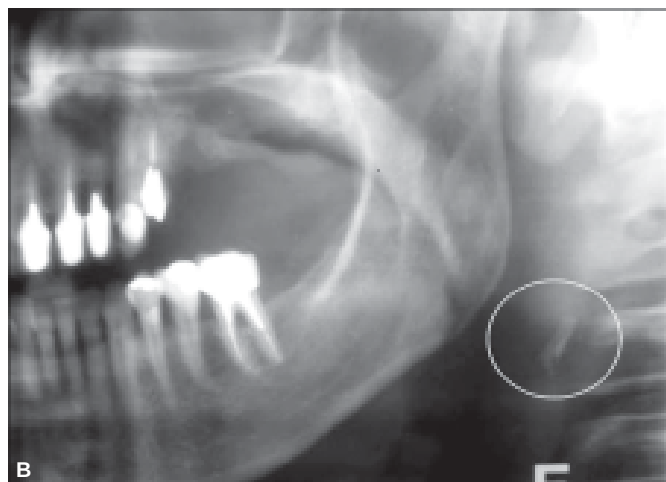
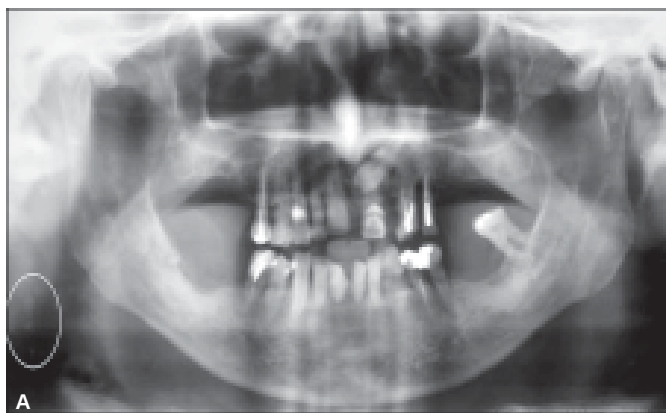
- *Inflammatory process*—it is sequelae of inflammatory process affecting the wall.
- *Other causes*—it is also found in Sturge-Webber syndrome and diabetes mellitus.

Clinical Features

- Usually no clinical sign or symptoms develop.

Radiographic Features

- *Site*—calcification with the carotid artery are located in the soft tissue below the angle of the mandible and between the hyoid bone and the image of the cervical spine (**Figs 29-4A and B**).
- *Margins*—calcific deposits on wall of artery will outline the image of the artery.



Figs 29-4A and B: OPG showing calcification in the carotid artery (Courtesy Dr Fábio de Lima Cravo).

- *Appearance*—a pair of thin opaque lines that may have either a straight course or a tortuous path. They may appear as amorphous or punctuate calcifications.
- *Internal structure*—the calcified wall appear as radiopaque circle.
- *Stroke*—in patient who is suffering from diabetes mellitus and having calcification in the region of carotid artery leading to macrovascular atherosclerosis can cause stroke.

Diagnosis

- *Clinical diagnosis*—it is not possible to make clinical diagnosis.
- *Radiological diagnosis*—calcified deposit seen on walls of artery.

Differential Diagnosis

- *Other calcific deposits*—usually, the linear nature of the calcified arterial wall indicates the nature of this condition.

Management

- This disease requires no treatment. Only special care should be taken for stroke.

Idiopathic Calcification

The deposition of calcium in normal tissue despite normal serum calcium and phosphate level is referred to as 'Idiopathic calcification'.

Sialoliths

It is described in Chapter 26: Salivary Gland Disorders.

Phleboliths

These are thought to be formed in older thrombi in veins or hemangiomas with slow blood flow. The thrombus organizes into granulation tissue and occasionally mineralizes with the deposition of calcium phosphate and calcium carbonate.

Clinical Features

- *Appearance*—the involved soft tissue may be swollen or discolored by the presence of veins or a soft tissue hemangioma.
- *Pressure application*—applying pressure to the involved tissue should cause a blanching or change in color if the lesion is vascular in nature.

Radiological Features

- **Site**—they are most commonly found in the hemangiomas.
- **Margins and shape**—the shape is round or oval with a smooth periphery. If it is viewed from side resembles a straight or slightly curved sausage.
- **Internal structure**—it may be homogeneously radiopaque but more commonly has a appearance of laminations. A radiolucent center seen, which may represent the remaining patent portion of the vessels (**Fig. 29-5**).



Fig. 29-5: Phleboliths showing multiple oval radiopacities (Courtesy RS Kamikawa).

Diagnosis

- **Clinical diagnosis**—swelling swollen tissue over the vein may suspect the disease.
- **Radiological diagnosis**—round shaped radiopacity with radiolucent center will give clue to the diagnosis.

Differential Diagnosis

- **Sialoliths**—sialoliths occur single as compared to phleboliths which is usually multiple.

Management

- Treatment of underlying cause should be done.

Metastatic Calcification

In this, calcium salts are precipitated in previous undamaged tissue. This precipitation is due to an excess of blood calcium and occurs particularly in such diseases as hyperthyroidism which depletes the bone calcium and causes a high level of blood calcium. The deposits of calcium occur in the kidney, lungs, gastric mucosa and media of blood vessels.

Ossification of the Stylohyoid Ligament

Ossification of stylohyoid is common and when it is associated with discomfort it is called as '*Eagle's syndrome*'. Ossification usually extends downward from the base of the skull and commonly occurs bilaterally. Bone tissue forms within segments of the stylohyoid ligament.

Clinical Features

- **Palpation**—there is hard pointed structure over the tonsil.
- **Symptoms**—vague pain on swallowing, turning the head and opening the mouth. Patient may describe earache, headache, dizziness or transient syncope which is caused by elongated styloid process impinging on glossopharyngeal nerve.
- **Signs**—there may be visible swelling on the region of the angle of mandible (**Fig. 29-6A**).
- **Stylohyoid syndrome**—clinical finding without the history of neck trauma constitutes '*Stylohyoid syndrome*'.



Fig. 29-6A: Swelling seen in angle of mandible region due to ossification of stylohyoid ligament.

Radiographic Features

- **Site**—styloid process appears as long, lapping, thin, radiopaque process between ramus of mandible and mastoid process. Ossification of ligament roughly parallels the posterior border of the ramus (**Fig. 29-6B**).
- **Appearance**—it appears as long, tapering, thin radiopaque process that is thicker at base and projects downward and forward.
- **Size**—it varies about 0.5 to 2.5 cm in length.
- **Internal structure**—small ossification appear homogeneously radiopaque. As the ossification increases in length and width, the outer cortex of its become evident as radiopaque band at the periphery.

Diagnosis

- *Clinical diagnosis*—earache, headache, dizziness with swelling at angle of mandible will suspect this disease.
- *Radiological diagnosis*—elongated stylohyoid process is easily identified on the radiograph (Fig. 29-6C).

Management

- *Amputation*—amputation of stylohyoid process should be carried out.

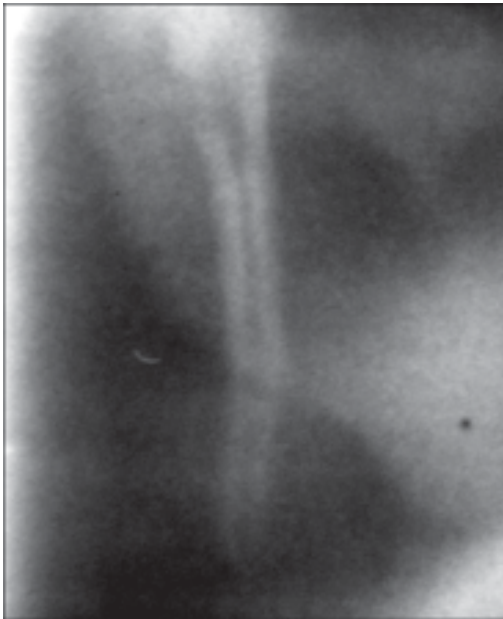


Fig. 29-6B: Calcified Outline of stylohyoid ligament (Courtesy RS Kamikawa).



Fig. 29-6C: Elongated styloid process with nodular mineralization pattern (Courtesy RS Kamikawa).

Osteoma Cutis

These are sites of normal bone formation in abnormal locations. The lesion occurs secondary to acne of long duration, developing in a scar or chronic inflammatory dermatosis.

Clinical Features

- *Site*—face is the most common site followed by lip (Fig. 29-7A) and tongue is the most common intraoral site.
- *Size*—size ranges from 0.1 mm to 5 cm and may be seen as single or multiple.
- *Color*—color may be normal or yellowish white.
- *Aspiration*—needle when inserted is met with stone-like resistance.



Fig. 29-7A: Osteoma cutis seen in lower lip region as slightly pale in color.

Radiographic Features

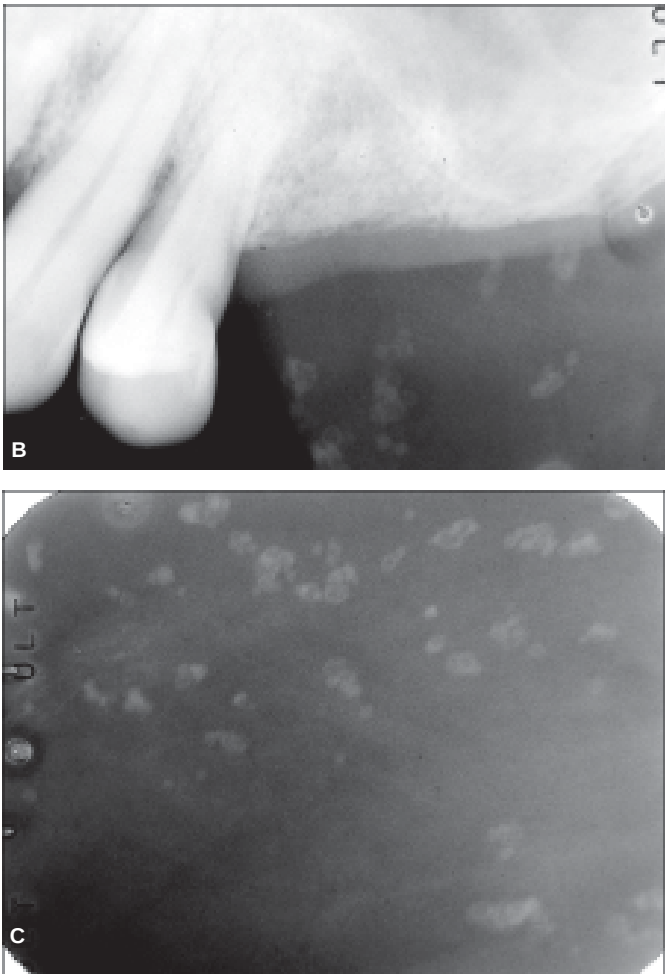
- *Site*—most commonly appears in the cheek and lip regions. Image can be superimposed over a tooth root or alveolar process.
- *Margins and shape*—smoothly outlined radiopaque washer-shaped image.
- *Size*—they are very small, although size can range from 0.1 to 5 cm.
- *Internal structure*—it may be homogenous radiopaque or may have a radiolucent center that represents normal fatty marrow.

Diagnosis

- *Clinical diagnosis*—There are areas of dense viable bone in the dermis or subcutaneous tissue.
- *Radiological diagnosis*—washer shaped image seen on the periapical radiograph (Figs 29-7B and C).

Differential Diagnosis

- *Myositis ossificans and calcinosis cutis*—osteoma cutis is more superficial than others.



Figs 29-7B and C: Multiple miliary osteoma cutis. Patient with history of large quantity of acne for a long period of time. Periapical radiographs showing evidence of ossifications in the soft tissues of the cheek (Courtesy RS Kamikawa).

Management

- **Surgical removal**—osteomas are occasionally removed for cosmetic reasons.

Myositis Ossificans

It is a condition in which fibrous tissue and heterotopic bone form within the interstitial tissue or muscle, as well as in associated tendons and ligaments. Secondary destruction and atrophy of the muscle occurs, as this fibrous tissue and bone interdigitate and separate the muscle fibers.

Types

- Localized myositis ossificans or traumatic myositis ossificans.
- Progressive myositis ossificans or generalized myositis ossificans.

Localized Myositis Ossificans

It is also called as 'Post-traumatic myositis ossificans' or 'Solitary myositis'.

Etiology

- **Trauma**—it is caused by acute or chronic trauma or heavy muscular strains caused by certain occupation or sports. Traumatization of the periosteum of an adjacent bone with the displacement of osteoblasts into the muscle and subsequent formation of bone.
- **Periosteal implants**—activation of periosteal implants already present in muscle by trauma or hemorrhage.
- **Metaplasia**—metaplasia of the pluripotential intermuscular connective tissue into the bone. Metaplasia of fibrocartilage.

Pathogenesis

- **Injury**—hemorrhage into the muscle or associated tendon or fascia → the hemorrhage organized and undergoes scarring → during healing process cartilage is formed → calcification of cartilage → ossification of cartilage.

Clinical Features

- **Age and sex**—it can occur at any age, sex and more often in young persons.
- **Site**—the most commonly involved muscles are the masseter and sternocleidomastoid but in some cases, lateral pterygoid muscle can be involved.
- **Symptoms**—site of trauma remains swollen, tender and painful much longer than expected. In some cases, there is a mild discomfort associated with a progressive limitation of motion.
- **Signs**—the overlying skin may be red and inflamed. Intramuscular mass is palpated at 2 to 3 weeks. The lesion may appear fixed or it may be freely movable on palpation.

Oral Manifestations

- **Site**—it involves the muscles of face particularly masseter and temporal following single traumatic injury.
- **Symptoms**—some difficulty in opening of the mouth.

Radiographic Features

- **Site**—radiolucent band can be seen between the area of ossification and adjacent bone and heterotopic bone may lie along the long axis of the muscle.
- **Internal structure**—faintly homogenous opacity. Delicate lacy or feathery internal structure of increased radio-density develop indicating bone has formed (**Fig. 29-8**). Sometimes it is accompanied by circumscribed cortical periphery.



Fig. 29-8: Myositis ossificans. See the increased radiodensity with margins are more radiopaque than internal structure (Courtesy Dr Bhaskar Patle).

- **Margins**—margins are more radiopaque than the internal structure.
- **Shape**—there is variation in shape from irregular oval to linear streaks running in the same direction as the normal muscle fibers. These *pseudotrabeculae* are characteristic of myositis ossificans and strongly imply a diagnosis.
- It lies close to the surface of the bone. With the passage of time the new bone is resorbed.

Diagnosis

- **Clinical diagnosis**—palpable intramuscular mass with swelling area of trauma may give clue to the diagnosis.
- **Radiological diagnosis**—pseudotrabeculae and more radiopaque margin is present.
- **Laboratory diagnosis**—biopsy shows degeneration of muscle and connective tissue hyperplasia to chondrification and ossification. The trabecular pattern is often extremely bizarre with the cartilage and myxomatous tissue present which may resemble callus formation.

Differential Diagnosis

- **Ossification of stylohyoid ligament, dystrophic calcification in areas of necrosis, pathological calcification and phlebitis**—the form and location of myositis ossificans are enough make the differential diagnosis.

Management

- **Rest**—sufficient rest should be given with limitation of use.
- **Excision**—excision after process becomes stationary.

Progressive Myositis Ossificans

It is characterized by formation of bone in tendons and fascia with subsequent replacement of adjacent muscle by expanded bony mass. In some cases, there is history of hereditary and familial pattern.

Clinical Features

- **Age and sex**—it usually affects children before 6 years of age. It is seen more in males as compared to females.
- **Progress**—it may advance rapidly or there may be long period of relative inactivity with intermittent bursts of activity.
- **Site**—starts in muscles of neck and upper back and moves to extremities.
- **Symptoms**—soft tissue swelling that is tender and painful and may show redness and heat.
- **Sign**—gradual increase in stiffness and limitation of motion of neck, chest and back and extremities. Ultimately entire groups of muscles become transformed into bone resulting in limitation of movements.
- **Associated anomalies**—it is associated with congenital small first metatarsal and metacarpal bones, small little bone. Interphalangeal joint may be fused.
- **Facial finding**—the masseter muscle is frequently involved so that fixation of jaw occurs.
- **Petrified man**—the patient becomes rigid and called as 'Petrified man'.
- **Prognosis**—patient dies during 3rd or 4th decades. Premature death is usually results from respiratory embarrassment.

Radiographic Features

- **Appearance**—there is evidence of dense osseous replacement of the greater part or whole of the muscle.
- **Internal structure**—densities of heterotrophic bone vary widely. The bone that is laid down in the muscle does not show structure of normal bone but it is rather structureless mass of variable density.
- **Linear striae of increase density**—coarse linear striae of increased density represent new bone formation in some cases; the streaks follow the long axis of the particular muscle involved. In early stages calcified deposits are granular and fragmentary.
- **Margins**—lesion have smooth or irregular margins, lying in close relationship with the bone.

- *Advanced lesion*—the dense masses with passage of time tend to coalesce. Skeleton becomes osteoporotic because of lack of function as muscles atrophy and joints become ankylosed.

Diagnosis

- *Clinical diagnosis*—‘Petrified man’ appearance with gradual limitation of movement of the body.
- *Radiological diagnosis*—bone in muscle show structureless mass of variable density.
- *Laboratory diagnosis*—the muscle in this disease is gradually replaced by connective tissue which undergoes osteoid formation and subsequently ossification. In some cases, cartilage formation may become evident.

Differential Diagnosis

- *Rheumatoid arthritis*—in initial stage, it is difficult but as disease progress specific anomalies confirms the diagnosis.
- *Calcinosis*—the deposits of amorphous calcium salts frequently resorb, but in progressive myositis ossificans the bone never disappear.

Management

- No effective treatment exists. Nodules that are traumatized and then ulcerated frequently should be excised.

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Section **4**

Systemic Diseases Manifested in Jaw



Introduction

The literal meaning of the word 'disease' is 'loss of ease'. The oral cavity reflects the state of systemic health more frequently than other parts of the body. Even in ancient time, examination of mouth and tongue was given great importance. The oral tissues are in direct physical continuity with rest of the body and they are also related via blood, lymphatics and nerve pathways. Furthermore, systemic influence such as endocrinological, immunological and psychological factors has an important role in the balance between oral health and disease.

During both, development and maintenance, local and systemic factors are concerned in disease process of mouth. Oral health must be considered in relation to general health. The dentist's role in general health is based on the fact that, he is the first person to see the oral lesion. In preventive dentistry, the utmost principle is of early diagnosis. Dentist has an important role in preventive medicine as many systemic diseases have primary oral manifestations.

Infections Caused by Bacteria

The following diseases are caused by bacteria.

Syphilis

It is also called as 'lues'. The incidence of syphilis is decreased after introduction of penicillin in late 1940s. Patient infected with *Treponema pallidum* and HIV may exhibit a malignant form of syphilis with slow development of standard serological response to syphilis. The tertiary manifestations lead to considerable morbidity and mortality.

Etiology

- *Sexual contact*—it occurs most exclusively by venereal contact.
- *Maternal transmission*—infection may transfer from mother to fetus.
- *Drug user*—incidence of syphilis is increased nowadays in crack cocaine abuse and barter of illegal drugs for sex.
- *Predisposing factors*—overcrowded living and primitive housing conditions are the predisposing factors for syphilis.

Classification

- *Acquired syphilis*—contacted primarily as venereal disease due to sexual intercourse with infected partner.
 - *Primary*—it is evident clinically 3 to 90 days after the exposure.
 - *Secondary*—it is discovered 4 to 10 weeks after primary stage.
 - *Tertiary*—this stage develop after the latent phase.
 - *Quaternary syphilis*—the atypical malignant progression of tertiary neurosyphilis in immunocompromised HIV individuals is referred as quaternary syphilis.
- *Latent phase*—this appear after secondary stage and in this stage, there is no sign and symptoms are present. Serological test is positive in this stage.
- *Congenital*—this is secondary to fetal infection.
- *Early syphilis*—primary syphilis, secondary syphilis and the early latent phase of the disease are grouped as early syphilis. Early syphilis may last up to two years and is infectious.
- *Late syphilis*—while late latent and tertiary are grouped as late syphilis. Late syphilis is locally destructive and non-infectious.

Primary Syphilis

Clinical features

- **Incubation period**—lesion develop at the site of inoculation approximately 3 to 90 days after the inoculation.
- **Site**—it occurs most frequently on penis in males and vulva or cervix in females. Recently, occurrence on extragenital sites have increased as a result of increase in orogenital sex and increased contact among the infected homosexuals. Extragenital sites of involvement include fingers, perianal region, nipples, lips, tonsils and intraoral structures such as tongue and palate.
- **Chancre**—it is slightly raised, ulcerated, non-tender, non-bleeding, firm plaque which is usually round, indurated and with rolled raised edges.
- **Size**—it varies in size from 5 mm to several centimeters.
- **Symptoms**—it is painless, unless superinfected. It disappears without therapy after 10 days.
- **Lymph nodes**—regional lymph nodes become firm enlarged, rubbery in consistency and non-tender.

Oral manifestations

- **Site**—oral lesions of primary syphilis are rare and occur at the site of entry of treponema. Chancre has been described on lips, oral mucosa, lateral surface of tongue, soft palate, tonsillar area, pharyngeal area and gingiva.
- **Transmission**—transmission can occur during kissing as a consequence of sexual practice among homosexual and heterosexuals, or by contact with objects such as mouth piece of musical instruments and medical or dental instruments.
- **Appearance**—it has narrow copper colored, slightly raised borders with reddish brown base in center.
- **Symptoms**—intraoral chancres are slightly painful due to secondary infection and are covered with grayish white film.
- **Size**—it measures from 0.5 to 2 cm in diameter.
- **Signs**—occasionally, it retains white sloughy material. In some cases, there is proliferation that resembles pyogenic granuloma.
- **Tonsils**—primary involvement of tonsils is manifested by considerable edema, redness, ulcerated and eroded lesion.
- **Lymph nodes**—regional lymphadenopathy occurs.
- **Extraoral lip chancre**—extraoral lip chancre may have more typical brown crusted appearance which may be multiple. Lower lip involved more frequently (**Fig. 30-1**).
- **Healing**—oral chancre heals spontaneously in 3 to 8 weeks leaving small scars.

Secondary Syphilis

Clinical features

- **Spread**—organisms proliferate and spread by the way of bloodstream to produce lesions elsewhere.



Fig. 30-1: Chancre present on the lower lip in male patient which has got slightly raised border (Courtesy Dr Jitu Sachdeo).

- **Incubation period**—it usually appears within 4 to 10 weeks after primary lesion.
- **Symptoms**—fever and generalized lymphadenopathy, which is painless, discrete and non-adherent to the surrounding tissues, may be seen. Headache, anorexia, pain in joints and muscles also occurs.
- **Skin**—when appear on skin, they manifest as fine macular or papular rash, sometimes accompanied by alopecia. The skin rash may resolve completely or leave residual areas of hypo- or hyperpigmentation.
- **Face**—circinate lesions on face are characteristic of secondary syphilis.
- **Mucus patches**—mucus patches are small, smooth, erythematous areas or superficial grayish erosions found on mucus membrane of vulva, penis, or in oral cavity, on palate and tonsils. They are described as snail track ulcers.
- **Condyloma latum**—condyloma latum are grayish, moist, flat topped, extra large plaque which sometimes coalesce into larger plaques, found on moist mucocutaneous surfaces such as vulva, anus, scrotum, thigh and axilla.
- **Split papule**—split papule is a double papule which occurs at skin folds and angle of mouth.
- **Lues maligna**—this is explosive and widespread form occurs in compromised immune system. It is characterized by formation of necrotic ulceration.
- **Lymph nodes**—generalized symmetrical enlargement of the lymph nodes in posterior cervical, suboccipital, supratrochlear and inguinal regions.
- **Latent phase**—patient may enter the phase of latency without treatment.

Oral manifestations

- **Mucus patches**—it is mucous membrane analogue of papular or macular skin eruptions.
 - **Site**—it is found on tongue, buccal mucosa, tonsillar and pharyngeal region, and lips.

- *Appearance*—it appears as slightly raised grayish white lesions surrounded by erythematous base.
- *Surface*—they are covered by grayish white membrane. Trauma results in raw bleeding surface.
- *Snail track ulcers*—confluence and coalescence of these glistening mucous patches gives rise to the so called 'snail track ulcers'.
- *Symptoms*—it is often painless but mild to moderately painful.
- *Split papule*—they are highly infectious.
 - *Site*—split papule is a raised papular lesion developed at the commissure of lip and with a fissure separating the upper lip portion from lower lip portion.
 - *Appearance*—they are called as split papules as they are cracked in the middle giving a 'split pea appearance'.
- *Condyloma latum*—they are flat silver gray wart like papule, sometimes having ulcerated surface. They are painless. Regional lymphadenopathy is usually present.

Tertiary Syphilis

Clinical features

- *Incubation period*—it may occur at any age from the third year upto the patient's life.
- *Forms*—in tertiary syphilis, 1/3rd develop benign or gummatous form, 1/3rd cardiovascular form and 1/3rd neurosyphilis, i.e. general paresis and tabes dorsalis.
- *Gumma*—gumma is due to a chronic destructive granulomatous process which occurs anywhere in the body. Gumma is the result of hypersensitivity reaction between hyperergic host and treponema. Single cerebral gumma may produce symptoms suggestive of brain tumor
 - *Types*—there are two types of gumma, i.e. central and cortical.
 - *Appearance*—the characteristic gumma appear as chronic granulomatous localized lesion, which ulcerates. It is nodular in appearance.
 - *Signs*—punched out ulcer with vertical walls and dull red granulomatous base is the typical clinical feature of ulcerative gummatous lesion.
 - *Cutaneous lesions*—cutaneous lesions heal slowly and leave behind tissue paper-like scars.
- *Neurosyphilis*—it occurs due to obliteration of small vessel artery involving vasa vasorum of aorta and other large vessels of the central nervous system (neurosyphilis). Neurosyphilis is manifested as tabes dorsalis and general paresis. Tabes dorsalis is the syphilitic involvement of dorsal column of spinal cord and dorsal root ganglion. General paresis is syphilitic involvement of cerebral tissue.
- *Tabes dorsalis*
 - *Symptoms*—patient loses the positional sense of his lower extremities and walks with a slapping step. Burning and pricking sensation of the extremities, paresthesia, or actual anesthesia of the part may accompany the characteristic gait.
 - *Positive Romberg's sign*—person is unable to stand erect unaided with his eyes closed.
 - *Tabetic crises*—short, shooting, knife-like pains may be experienced in the abdominal region called 'tabetic crises', which results from involvement of the dorsal root ganglion.
 - *Charcot's joint*—trophic changes consist of deep perforating ulcers and painless destruction of larger joints.
- *General paresis*
 - *Argyll Robertson pupil*—pupils that react to accommodation but not to light.
 - *Symptoms*—increased irritability, fatigue, mental sluggishness and carelessness in personal habits.
 - *Signs*—loss of fine muscular coordination is indicated by inability to enunciate clearly or to perform delicate tasks with the hands.
 - *Spinal cord involvement*—involvement of spinal cord is late manifestation characterized by paresthesia, burning and prickling sensation in the extremities.
 - Patient may get unrealistic ideas of wealth or ability.
- *Cardiovascular syphilis*—it occurs in 10% cases of late syphilis. Involvement of CVS in tertiary syphilis affects aorta and aortic valve and 80% of deaths occur due to it. There is medial necrosis and destruction of elastic tissue occurs in the wall of large blood vessels. Dilatation and aneurysm occurs.
- *Tertiary syphilis in HIV patient*—a recent feature has been an apparent alteration in the behavior of syphilis in HIV positive patients. Natural history of syphilis may be altered in HIV positive patients and this group is at high risk for the development of tertiary syphilis. Genital ulcerations due to syphilitic infection allows a portal of entry for the viral particles and may lead to greater risk of infections in these patients.

Oral manifestations

- *Site*—gumma can occur anywhere in the jaw but are more frequently on palate, mandible, and tongue.
- *Gumma*
 - *Appearance*—gumma may manifest as solitary, deep, punched out mucosal ulcer.
 - *Symptoms*—breathing and swallowing difficulty may be encountered by the patient.
 - *Progress*—it usually starts as small, pale, raised, nodular mass in the midline of the palate which ulcerates and rapidly progresses to the zone of necrosis.

- *Signs*—it may cause perforation of palatal vault.
- *Punch out defects*—lesion is sharply demarcated and the necrotic tissue at the base of the ulcer may slough away leaving punched-out defects.
- *Tongue*—numerous small healed gummata in tongue results in series of nodules or scars in deeper areas of the organ, giving the tongue an *upholstered or tufted appearance*.
- *Leutic glossitis*—complete atrophy of papillary coating and firm fibrous texture seen. It is called as leutic glossitis. Initially, it is thought to be precancerous but nowadays, this concept is disputed. Loss of papillae is probably due to endarteritis leading to circulatory deficiency of lingual vasculature.
- *Chronic superficial interstitial glossitis*—the tongue may involve diffusely with gumma and appear large, lobulated and irregularly shaped. This lobulated pattern is called as chronic superficial interstitial glossitis.

Radiographic Features of Syphilis

Bones are not known to be involved in the primary stage. Bone involvement usually occurs in tertiary stage and in some cases, in secondary stages.

Gummatous Lesion of the Bone

- *Site*—skull is one of the usual sites of gummatous involvement frequently involving frontal, parietal, and temporal bone. Maxilla is affected more than the mandible, the common site being the hard palate particularly the central portion. It begins as extension of gumma from the palatal mucosal aspect or from the floor of nose. Multiple gummas of oral structure may invade the coronoid process and medial pterygoid muscle.
- *Central gumma*—there is radiolucent area with ill-defined margins which merge with the normal bone or there are multiple areas of greater radiolucency. These areas are separated from one another by normal looking bone. With the passage of time, radiolucent areas are fused and appear as one dark shadow which is caused by perforation of the palate.
- *Cortical gumma*—it starts on the surface of bone and destroys a wide area of the cortical covering. The deeper cancellous bone is involved later and destroyed. The result is that there is a well-marked area of bone destruction, open to the surface of the bone and has sharply defined margins, which are devoid of rarefaction.
- *Diffuse gummatous infiltration*—it is usually found in the vault of the skull in the vicinity of the intramedullary gummata. Mandible is involved more often than maxilla. Suppuration and sequestration are always present. Radiographic appearance suggests widespread

osteomyelitis which has not been treated. There are many islands of bone with widely varying shape and size, many with grossly irregular borders, which are well defined. The density of the islands of bone varies from normal bone to rarefied bone.

Syphilitic Periostitis

It is the most common manifestation of congenital syphilis and may also be seen in secondary stage of acquired syphilis. The lesion may develop very late in the disease process and is characterized by periosteal reaction, which may either be limited and circumscribed or extensive.

- *Site*—it involves mandible more commonly and it appears as a single layer or several layers of new bone, more or less parallel with the margins of jaw. The calvarium is one of the usual sites of involvement in this manifestation of syphilis.
- *Network of lattice*—a rare manifestation of syphilitic periostitis is new bone arranged with trabeculae forming a loose type of pattern which is best described as a gross caricature of a network of lattice. There is often a dark line between the new bone and jaw and between the layers of new bone.
- *Syphilitic osteitis*—there are small superficial areas of rarefaction, in the underlying bone, due to syphilitic osteitis.

Congenital Syphilis

It is infection of fetus established by the passage of spirochetes from mother, through the placenta. Transplacental infection after 18-week gestation is related to development of immune complement rather than any toxic effect on organism.

Congenital syphilis has got three diagnostic features called as *Hutchinson's triad* which includes hypoplasia of permanent incisors and 1st permanent molars, eighth nerve deafness and interstitial keratitis.

Clinical features

- *Early manifestations*—it is manifested within the first 2 years of life (neonatal congenital syphilis) as rhinitis and chronic nasal discharge with maculopapular eruptions, other than mucocutaneous lesion and loss of weight. The lesions can be seen in spleen, kidney, bones and CNS. Bullae, vesicle and superficial desquamation with cracking and scaling of reddened soles and palms, petechiae, mucus patches and condyloma latum also occurs.
- *Late manifestations*—after 2 years, interstitial keratitis (opacification of corneal surface with resultant loss of vision), vascularization of cornea, eighth nerve deafness, arthropathy, signs of congenital neurosyphilis, gummatous destruction of palate and nasal septum develop. There are also saber shins or anterior tibial bowing.

- *Higoumenakis's sign*—irregular thickening of sterno-clavicular portion of clavicle.

Oral manifestations

- *Postrhagadic scarring and syphilitic rhagades*—post-rhagadic scars are linear lesions found around oral and anal orifices. They result from diffuse leucic involvement of the skin in these areas from 3rd to 7th week after birth. They appear as red or copper colored linear areas covered with a soft crust. Rhagades are said to be more frequent on the lower lip. Healed syphilitic rhagades appear as ordinary cicatrices.
- *Linear scars*—the linear scars are radially arranged and perpendicular to the mucocutaneous junction, which are more prominent on lower lip near angle of mouth.
- *Changes in dentition*—retarded root resorption of deciduous dentition. There may be 'marring' of permanent incisors present in congenital syphilis. 6 to 28% of the incisors and 3 to 37% of the molars have hypoplasia. Spacing between cuspid and incisors is present. Malocclusion and open bite is present.
- *Molar features*—the crown of the first molar in congenital syphilis is irregular and enamel of the occlusal surface and occlusal third of the tooth appears to be arranged in agglomerate mass of globules, rather than in well formed cusp.
- *Screw driver shaped incisors*—constriction of crown toward incisal edge screw results in driver or peg shaped incisor. In addition, incisal edge is usually notch which may be due to the absence of central tubercle or calcification center. Rounding of mesial and distal incisal line angles.
- *Moon molars*—in molars, positioning of the cusps toward the central portion of the crown, gives the tooth a bud shaped or a shrunken occlusal form called as mulberry molars or Moon's molars. Affected molars are dirty yellow in color due to hypocalcification.
- *Carabelli cusp*—prominent accessory mesiolingual cusp of upper molar (Carabelli cusp).
- *Jaw bones*—defective maxilla which is hypoplastic and short with relative mandibular prognathism. Frontal bossing and saddle nose deformity occurs.
- *Lesion biopsy*—histopathological examination of suspected lesion, stained by silver impregnation technique, is useful particularly when the lesion contains few organisms, as may be in case of tertiary lesion. For oral lesions, this technique is of considerable value.
- *Treponemal antigen test*—Treponemal test is of value in making a confirmatory diagnosis while non-treponemal test is of value in assessing the efficacy of the therapy.
- *Non-treponemal (antigen) test*—commonly used non-treponemal test is Venereal Disease Research Laboratory (VDRL) and rapid plasma reagin (RPR) test. Both tests are inexpensive, simple and rapid to perform but require competent experienced laboratory to minimize the risk of false positive or false negative results.

Management

- *Antibiotics*—patient should give benzathine penicillin (2.4 million units IM) aqueous crystalline penicillin, tetracycline hydrochloride (500 mg orally 4 times a day for 15 days). Patients who are allergic to penicillin erythromycin (500 mg by mouth 4 times a day for 15 days). *T pallidum* disappears from infectious lesion within 24 hours of instituting therapy.
- *Follow up*—patient should be followed with repeated physical examination and repeated VDRL is to be done at 1, 3, 6, 9, 12, 18 and 24 months. At the end of 24 months, if VDRL is negative, patient is said to be cured.
- *Prevention*—there is no dependable prophylactic measure other than sexual abstinence with infected partner. The use of prophylactic antibiotics locally is beneficial in pregnant women suspected to exposure. Prevention of congenital syphilis can be achieved by subjecting pregnant women to antenatal and postnatal check up.

Non-venereal Treponematoses

Endemic syphilis is also called as 'bejel' which is caused by spirillar form of *Treponema palladium*, yaws which is caused by *Treponema pertenue* and pinta which is caused by *Treponema carateum* are most non-venereal treponematoses which occur in children and causes destructive skin and bone diseases. These diseases run a milder course compared to venereal treponematoses.

Endemic Syphilis

It is also called as 'bejel' and most commonly occurs in childhood.

Transmission

- *Direct transfer*—direct transfer is through lesion to skin contact.
- *Indirect transfer*—indirect transfer is through the common use of drinking, bowls, or possibly by flies.

Diagnosis of syphilis

The presence of clinical manifestations together with history of a sexually active person should give clue to the diagnosis of acquired syphilis.

- *Dark field examination microscopy*—It is the most useful method of identifying spirochete in primary acquired and occasionally, secondary syphilis. Not reliable for oral lesions, since the normal flora contains non-pathogenic treponema which are difficult to distinguish from *T. pallidum*.

Clinical features

- *Age*—the disease is primarily seen in children, although adult cases are often reported.
- *Early stage*—the early stage disease is characterized by the appearance of mucus patches in the oronasopharyngeal region, angular stomatitis, skin rashes, pigmentary changes and tenderness of the long bones and regional lymphadenopathy.
- *Oral patches*—oral mucus patches are shallow, relatively painless lesions, initially seen as white which soon become erosive. The lesions are usually found on the lips, tongue and other intraoral surface.
- *Skin lesion*—skin lesions may be papular, papillomatous, macular, papulosquamous, annular or circinate.
- *Sabre tibia*—osteoperiostitis usually involving long bones is the most common manifestation which may cause nocturnal pains in the legs. Long standing osteitis and periostitis may eventually lead to forward bowing of tibia known as 'sabre tibia'.
- *Late stage*—it is seen where early endemic syphilis remains untreated for 6-9 months or even longer as an alteration in about 1/3rd of the patients or as tertiary state of the disease. Late lesions are mainly of gummatous type, involving skin, mucous membrane and bone. Skin lesions are usually extensive, chronic, destructive, scarring and with depigmentation. Gummatous destruction of nasal septum is common.
- *Rhinopharyngitis mutilans*—destruction of the lips, soft palate and nasopharynx can occur leading to gross deformity of the face.

Diagnosis

- *Clinical diagnosis*—Rhinopharyngitis mutilans with sabre tibia will give clue to the diagnosis.

Management

- *Penicillin*—it is the drug of choice and where penicillin is contraindicated; drugs such as tetracycline and erythromycin can be given.
- *Surgical correction*—surgical correction of the defects of face should be carried out for aesthetic purpose.

Pinta

It is almost exclusively confined to the Western hemisphere. The mode of transmission is either by direct or indirect contact.

Clinical features

- *Incubation period*—the incubation period is usually 2-3 weeks, after which the disease is clinically manifested. Infection is seen in young adults of 15-30 years of age.
- *Subcutaneous lesion*—the basic lesion of pinta is a solely developing subcutaneous granulomatous lesion.

- *Primary lesion*—it is believed to occur at the point of contact. They begin as small erythematous papule which enlarges within a year or two. In early lesion, pigment is lost from the germinal layer of epidermis and may become concentrated in upper layer. The primary lesion may be found on trunk, leg, face and around the anus and may be surrounded by satellite lesions.
- *Pintides*—secondary lesion of pinta is known as pintides, which appear after a period of months of years. It starts as papular lesion and develops into plaques with scaly and centrally pigmented areas. These lesions tend to become eventually depigmented. Lesions are itchy and painless. Moderate lymph node enlargement is often seen mostly due to accompanying secondary infections.
- *Late lesion*—over the period of time primary and secondary lesions become depigmented and the skin over these lesions become atrophic. Hyperkeratosis of the palms and soles accompany the formation of the achromic lesion at this stage.

Diagnosis

- *Clinical diagnosis*—skin lesion with pintides and subcutaneous granulomatous lesion will give clue to the diagnosis.

Management

- Penicillin is the drug of choice. Patients who are allergic to penicillin, tetracycline can be given.

Yaws

It is also called as 'framboesia' or 'buba'. It is caused by *Treponema pertenue*, which is pathogenic for monkeys, rabbits and hamsters. It is transmitted either by direct contact with the exudates of the early infectious lesion or indirectly by contaminated utensils.

Clinical features

- *Primary stage*—the incubation period is between 9 to 90 days. The initial lesion, a papule, appears at the site of entry of the *Treponema*. It occurs usually through an abraded or lacerated site. The lesion may ulcerate and infection can spread via bloodstream. Regional lymph nodes are usually palpable. Children with only primary lesion may complain of pain.
- *Secondary stage*—usually 1 to 3 months after the appearance of the papular primary lesion, infection spreads and a painless papilloma or frambesial granuloma appears. Lesions in the secondary stage are usually found in warm and moist sites such as axilla, groin and the skin around the natural orifices, including the mouth. They may be multiple and condylomatous in appearance. Partially, healed skin lesions often have an annular or

circinate appearance. Skin lesions may spontaneously heal in about 3 to 6 months. Bone and cartilage are involved. Lesions include osteitis, periostitis and dactylitis. In infected growing children, 'sabre tibia' develop. Plantar and palmar hyperkeratosis is also seen.

- **Tertiary stage**—tertiary lesion may occur after 5 years of primary lesion. The stage is characterized by gummatous nodular ulcerative lesion. Gummata of the skin start as subcutaneous nodules which eventually ulcerate. Osteoperiostitis, area of rarefaction and necrosis occur in long bones.

Oral manifestation

- **Lips**—during secondary stage, mucosal surface of lips may involve due to direct extension of the lesion present at the mucocutaneous junction.
- **Goundou**—secondary changes of the nasal processes of maxillary bone have been reported during the secondary and tertiary stages of the disease. These result in thickening of the face on either side of the bridge of the nose, gives rise to a characteristic facial appearance called 'goundou'.
- **Gangosa or saddle nose defect**—in some cases, the lesion starting either on the soft palate, uvula or hard palate, eventually invades and destroys soft and bony parts of nose which is called as 'Gangosa' thereby causing 'saddle nose defect'.

Diagnosis

- **Clinical diagnosis**—saddle nose defect with sabre tibia goundou shape appearance gives clue to diagnosis.

Management

- **Penicillin**—it is the drug of choice.

Gonorrhea

It is primarily an infection of the genitourinary tract mucosa. It is caused by gram -ve intrabacillary located diplococcus *Neisseria gonorrhoeae*. It is an oval, paired, gram-negative microorganism.

Local infection may occur at extra-genital site (Rectal and pharyngeal gonorrhea). Rectal mucosa is affected in 30% to 50% of women with urogenital gonorrhea. Uncomplicated local infection at other extra-genital sites is rare in adults.

Early sexual awakening, prostitution and varied sexuality are believed to be responsible for the increase incidence of gonorrhea.

Pathogenesis

- **Penetration of gonococci**—once the gonococci are directly deposited on the genitourinary tract mucosa during

sexual intercourse, they penetrate through the inter-cellular spaces of the epithelium and reach the subepithelial connective tissue.

- **Inflammatory reaction**—within 2 to 3 days of infection, an intense inflammatory reaction occurs resulting in characteristic mucopurulent discharge through urethral lumen.
- **Chronic stage**—a chronic stage may be reached, if untreated and spread is either by direct extension through lymphatics or hematogenous route.

Clinical Features

- **Age and sex**—it is primarily a disease of young adults between the ages of 15 to 24 years and is more common in males, as compared to females.
- **Location**—it is seen on genital site. Occasionally, it can involve extra-genital sites. In male, urethra is involved and in females, cervix is involved.
- **Incubation period**—incubation period is 2 to 5 days.
- **Symptoms**—there is profuse purulent urethral discharge with frequent micturition, followed by dysuria. In some of the patients, there may be fever and headache.
- **Gonococcal septicemia**—features of this include myalgia, arthralgia, polyarthrititis, dermatitis, fever, endocarditis, meningitis and oral manifestation.
- **Skin lesion**—dermatologic manifestation includes discrete papules and pustule that often contain of hemorrhagic component.
- **Gonococcal ophthalmia neonatorum**—this is the infection of infant eye which is transferred from infected mother. It may cause blindness.
- **Complication**—Cooper abscess (inflammation of periurethral glands), urethral stricture (difficulty in passing urine), arthritis, meningitis, endocarditis, epididymitis (painful swollen testicle), salpingitis or pelvic inflammatory disease (lower abdominal pain, metrorrhagia and pelvic tenderness on vaginal or rectal examination).
- **Diagnosis**—in all forms of it, including those of oral cavity and pharynx, the diagnosis rests on the identification of organism. Methods which are used include, gram stained smear, culture studies and direct fluorescent antibody test.

Oral Manifestations

- **Pharyngeal gonorrhea**—Pharyngeal gonorrhea is a term used for patients in whom *neisseria gonorrhoea* is isolated from nasopharynx. It is higher in pregnant women, sexually active homosexuals and heterosexuals practicing oral sex. History of fellatio is more associated with pharyngeal gonorrhea. Patient also noticed sore throat and evidence of pharyngitis.

- *Gonococcal stomatitis*—incidence is very rare and often shows multiple, painful and round elevated gray white eroded spots, with or without pseudomembrane formation. Regional lymphadenopathy may be seen. The wide range of lesion may develop in gonococcal stomatitis i.e. isolated ulcers, gingivitis and membranous gingivostomatitis.
- *Gingivitis*—acute gingivitis develops around extraction site in patient who practices fellatio repeatedly for days after extensive dental extraction. Gingiva may become erythematous, with or without necrosis.
- *Lips*—lips may develop acute painful ulcerations, limiting the motion.
- *Tongue*—the tongue may present red, dry ulcerations or become glazed and swollen with painful erosion with similar lesions on buccal mucosa and palate.
- *Temporomandibular joint involvement*—gonococcus infection involving articulating joint is the most common form of extragenital gonorrhea. Any joint may be affected, the commonest being knee, ankle and wrist. TMJ is affected in 14% of patients. It occurs as a result of hematogenous spread. Patient noticed difficulty in jaw movements due to pain and swelling of single or both joints is the presenting symptom. Rarely, perforation to the tympanic plate occurs. It may lead to fibrous ankylosis because articular cartilage is destroyed.
- *Types of gonococcal arthritis*—Wright has divided Gonococcal arthritis in two different categories:
 - *Category I*—Definite gonococcal arthritis—gonococci recovered in an aspirate from the affected joint.
 - *Category II*—Probable gonococcal arthritis. No gonococci recovered from the joint, but each of the following conditions exist.
 - Presence of gonococcal urethritis proven on smear or culture.
 - Arthritis occurring within 3 weeks of probable gonococcal infection.
 - Articular manifestations responding rapidly to antigenococcal therapy.

Radiographic Features

Radiologically, the joint reveals no abnormalities. Rarely, perforation through the tympanic plate may occur.

Diagnosis

- *Clinical diagnosis*—purulent urethral discharge with pharyngitis with TMJ involvement can give clue to diagnosis.

Differential Diagnosis

- *Primary syphilitic lesion*—painless indurated edema, painless lymph node swelling, causative agent in lesion.

- *Tuberculosis ulcer*—undermined, flabby border—usually painless, tuberculin test positive.

Management

- *Antibiotics*—single dose of the broad-spectrum cephalosporin antibiotic ceftriaxone 125 to 250 IM plus doxycycline 100 mg orally, twice a day for 7 days. In case of patient allergic to above drug, sepectinomycin 2 gm IM plus doxycycline. In some cases, orally or intramuscular administered fluorinated Quinolones may be helpful.
- *Prophylactic treatment*—prophylactic ophthalmic erythromycin should be given to prevent gonococcal ophthalmia neonatorum.
 - Recurrence—without adequate treatment, disease will recur.

Streptococcal Tonsillitis and Pharyngitis

The most common cause of this condition is beta hemolytic streptococci, adenoviruses, enteroviruses, influenza and parainfluenza.

Clinical Features

- *Symptoms*—sudden onset of sore throat, fever, dysphagia.
- *Signs*—redness of oropharynx and tonsils (**Fig. 30-2**), palatal petechiae and yellowish tonsillar exudate.
- *Lymph nodes*—there is cervical lymphadenopathy.
- *Systemic features*—headache, anorexia, abdominal pain and vomiting can also occur.



Fig. 30-2: Inflammation of pharynx and tonsil seen in streptococcal pharyngitis and tonsillitis.

Diagnosis

- *Clinical diagnosis*—tonsillitis with cervical lymphadenopathy will lead to diagnosis.

Management

- *Antibiotics*—penicillin and cephalosporin should be given. Erythromycin should be used in patient who is sensitive to penicillin.
- *Analgesic*—analgesic should be given to control pain and inflammation.
- *Warm saline gargle*—it is also effective treatment in case of tonsillitis.

Leprosy (Hansen Disease)

It is a chronic infectious disease which has predilection for the skin, nerves and mucous membrane. It probably originated in tropic and spread to the east. Leprosy has always been considered in superstitious dread and the person suffering from leprosy was considered unclean and socially outcasted.

It is caused by the leprae bacillus, *Mycobacterium leprae*, first observed by Hansen in 1868. It is not been possible to grow the bacillus in culture media. It is an acid fast, gram positive, non-motile bacteria with affinity for Schwann cells and cells of reticuloendothelial system.

Pathogenesis

- *Multiplication of bacteria*—after entry in the body the bacilli reach the lymphatic and bloodstream and are taken up by Schwann cells in peripheral nervous system, where they start multiplying. If the host cell mediated immunity is adequate, bacilli are destroyed and there is no disease. In the host, immunity is unstable and suboptimal, there will be some restricted multiplication of bacilli and lesion will develop.
- *Effect of immunity*—the variable status of host cell mediated immunity is reflected in the different clinical types of leprosy. When there is relatively good immunity but not enough to eliminate the infection, a localized type of disease called as tuberculoid type, is seen. When the host cell immunity is deficient a generalized form of the disease lepromatous leprosy develops.
- *Borderline variety*—in between these two polar varieties of the disease, there is a wide spectrum of manifestations, categorized as borderline leprosy.

Types

- *Tuberculoid leprosy*—it develops in patient with high immune reaction. It is localized. It is a benign form of leprosy involving the skin, nerves and regional lymph node.

- *Lepromatous leprosy*—this is present in reduce cell mediated immune response.
- *Borderline leprosy*—it can be tuberculoid borderline or lepromatous borderline.
- *Polyneuritic*—in this, multiple nerves are involved by the bacteria.
- *Indeterminate*—no specific lesion is found in this type of leprosy.
- *Erythema nodosum leprosum*—this is the severe form of leprosy.
- *Paucibacillary leprosy*—it corresponds to tuberculoid type of leprosy and lesions are limited to skin.
- *Multibacillary leprosy*—this corresponds to lepromatous type and involves multiple organs.

Clinical Manifestations

Tuberculoid type or paucibacillary type

- *Incubation period*—incubation period of 2 to 5 years during which patient passes through silent or latent period.
- *Sex*—males are affected more commonly than females with ratio of 3:1.
- *Skin*—lesions are hypopigmented, erythematous and flat or raised cutaneous lesions. Nerve involvement with loss of different types of sensation is also manifested.
- *Early tuberculoid leprosy*—it is manifested by hypopigmented macules which are sharply demarcated and hyperesthetic.
- *Intermediate tuberculoid lesion*—later, the lesions are larger with elevated and circinate margin. There is peripheral spread (Fig. 30-3) and central healing. At the end of this stage, the symptoms are those of irritation of nerve ending in the skin, persistent or recurrent paresthesia and numbness localized to certain area with no accompanying visible alteration in the corresponding skin lesion.



Fig. 30-3: Lesion of tuberculoid leprosy showing peripheral spread (Courtesy Dr Pincha).

- **Fully develop lesion**—in this, lesions are densely anesthetic and loose normal skin organs (sweat glands and hair follicles). There may be severe neuritic pain. Loss of eyebrows and eyelashes are prominent features of later involvement.
- **Peripheral nerve**—the sequelae of peripheral nerve involvement may develop in some cases and this may give rise to muscle atrophy, like contracture of hands and feet, loss of phalanges, lagophthalmus, exposure keratitis and corneal ulceration leading to blindness.

Lepromatous type or multibacillary leprosy

- **Age and sex**—this form is more commonly seen in children and females are affected more as compared to males.
- **Site**—this malignant form of the disease produced widespread involvement of body skin, peripheral nerves, mucous membrane, lymph nodes, eyes, skeleton, testes and other internal organs. It runs chronic course and seldom causes sudden death.
- **Appearance**—it develops early as erythematous macules (Fig. 30-4) or papules without subsequently lead to progressive thickening of skin and the characteristic nodules.
- **Margins**—the borders of the lesion are ill defined and centers of the lesion are indurated and convex.
- **Eyes**—loss of lateral portion of eyebrows is common.
- **Lymph nodes**—painless inguinal and axillary lymphadenopathy is common along with sterility and gynecomastia.
- **Nerve involvement**—nerve involvement is a late phenomenon.



Fig. 30-4: Macular lesion seen on the leg of patient in lepromatous leprosy.

- **Claw hand**—this is one of the typical features of leprosy (Fig. 30-5).



Fig. 30-5: Claw hand seen in lepromatous leprosy.

Erythema nodosum leprosum (ENL)

- **Occurrence**—they occur in lepromatous and borderline lepromatous patients, most frequently in the latter half of the initial year of treatment.
- **Signs**—tender, inflamed subcutaneous nodules develop, usually in crops. Each nodule lasts a week or two, but new crop may appear.
- **Symptoms**—low grade fever, lymphadenopathy and arthralgia can accompany severe ENL.
- **Leporma**—this is common feature of erythema nodosum leprosum (Fig. 30-6).

Facial manifestations

- **Prevalence**—depending on the type and duration of the disease, all patients with lepromatous leprosy show facial and oral manifestations and it is rare in tuberculoid and borderline leprosy.
- **Leonine facies**—the skin of face and forehead become thickened and corrugated giving a patient distorted facial appearance (leonine facies) (Fig. 30-7).



Fig. 30-6: Leproma in erythema nodosum leprosum presented as inflammatory swelling.



Fig. 30-7: Facial appearance in leprosy (note the saddle nose).

- *Ear*—earlobe becomes pedunculous due to lepromatous infiltration of the helical and lobes of ear.
- *Symptoms*—there is nasal stiffness, epistaxis, hoarseness.
- *Appearance*—the lesions are macular or raised, well defined, hypopigmented, unhydropic and hyperaesthetic or anesthetic.
- *Advanced cases*—advanced cases are characterized by nodular involvement of facial, cutaneous and subcutaneous structure.
- *Neural changes*—peripheral nerves such as facial and trigeminal nerves are frequently involved. Facial paralysis is observed in 3% of cases, complete or partial facial paralysis may occur either unilaterally or bilaterally. Paralysis of the orbicularis muscle results in facial disfigurement, difficulties in phonation and mastication as well as drooling. Involvement of trigeminal nerve results in hypoaesthesia and anesthesia.
- *Facies leprosa*—rhinomaxillary changes termed facies leprosa consist of atrophy of anterior nasal spine, atrophy and recession of the maxillary alveolar process confined to the incisor region and endonasal inflammation.
- *Saddle nose*—from nasal mucosa, granulomatous tissue invade cartilage and bone resulting in nasal collapse. The appearance is called as saddle nose (Fig. 30-7).
- *Chronic osteomyelitis*—destruction of facial cartilage and bone is caused by direct infiltration and secondary infection which results in chronic osteomyelitis.

Oral manifestation

- *Prevalence*—it is more commonly seen in lepromatous
- *Site*—involvement of oral mucosa, pre-maxilla, soft palate, uvula, tongue, gingiva and periodontium has been reported.

- *Lepromas*—small tumor like masses called as lepromas develop on the tongue, lips or hard palate. These nodules have a tendency to break down and ulcerate. Continuous infection may lead to scarring and loss of tissue.
- *Gingiva*—gingival hyperplasia with loosening of teeth have been also reported.
- *Tooth size and shape*—in association with severe and granulomatous infiltration of pre-maxilla in childhood, the tooth diameter is suddenly and concentrically reduced, while the less marked cases exhibit a well demarcated, tapering and shortening of root.
- *Enamel hypoplasia*—transient interruption of odontogenesis results in slight, circumferential hypoplasia of enamel and cementum.
- *Pulpal necrosis*—in long standing lepromatous leprosy, invasion of the pulp by granulomatous tissue causes pulpal necrosis leading to a pinkish discoloration of crown. Anterior teeth are most commonly affected.

Diagnosis

- *Ziehl-Neelsen stain*—skin smears should be examined as a routine with Ziehl-Neelsen stain. The percentage of solid bacilli in a smear is known as morphological index.
- *Biopsy*—skin, mucosal and nerve biopsy for histopathological examination are helpful in doubtful cases.
- *Lepromin test*—lepromin test is non-specific test to determine the hypersensitivity reaction and is useful in determining the immunological status of patient for classification of leprosy.

Differential Diagnosis

- *Gummatous lesion of syphilis*—VDRL test should be performed.
- *Ulcerative proliferative lesion of coccidioidomycosis and sporotrichosis*—peripheral nerve involvement is common in leprosy.

Management

- *Paucibacillary leprosy*—it is treated with 6 month regimen of rifampin and dapsone. Patient allergic to rifampin are treated with clofazimine, ofloxacin and minocycline.
- *Multibacillary leprosy*—this is treated with 24 months therapy of rifampin, dapsone and clofazimine.
- *Reconstructive surgery*—reconstructive and plastic surgery is essential for deformities of the hand and face
- *Vaccine*—BCG vaccines provide some protection against non-lepromatous leprosy.
- *Social development*—environmental changes with social and economic development are useful.

Tuberculosis

It is a systemic infectious disease of worldwide prevalence and of varying clinical manifestations. It is an infectious granulomatous disease caused by acid-fast bacilli *Mycobacterium tuberculosis* or rarely, *Mycobacterium bovis*. It can rarely be transmitted through the placenta from the diseased mother to fetus.

Pathogenesis

- **Primary infection**—initial tuberculosis usually occurs in lungs but occasionally, occurs in tonsil or alimentary tract. The common cause of primary infection is inhalation. In most of the patients of primary infection, the associated lymph node lesions heal and calcify.
- **Rupture of caseous tuberculosis foci**—in some cases, caseous tuberculosis focus ruptures into vein and produces acute dissemination throughout the body, a condition called as acute miliary tuberculosis. Meningitis often complicates this condition.
- **Progressive pulmonary tuberculosis**—progressive pulmonary tuberculosis may develop directly from a primary lesion or may occur following reaction of an incompletely healed primary focus.
- **Post-primary pulmonary tuberculosis**—post-primary pulmonary tuberculosis is a condition in which, there is a liquefied center of tuberculosis, pulmonary infection is discharged into sinus. Extension of infection into pleura causes tuberculosis pleurisy.

Etiology

- **Causative organism**—the first member identified as tuberculous bacillus and designate as *Mycobacterium tuberculosis*. Other microorganisms associated are *Mycobacterium bovis*, *Mycobacterium kanasasi*, *Mycobacterium xenopi* and *Mycobacterium malmoeense*. The organism is anaerobic, non-motile, non-sporing, rod shaped and is stained with special Ziehl-Neelsen stain.
- **Constitutional factor**—it is more common in low income group, unhygienic living conditions, malnutrition and overcrowding.

Predisposing Factors for Oral Infection

- **Systemic factors**—systemic factors like lowered host resistance and increased virulence of the organism may lead to oral manifestation in the tuberculosis patient.
- **Local factors**—local factors like poor oral hygiene, local trauma, presence of pre-existing lesion such as leukoplakia, periapical granuloma, dental cyst, dental abscess, jaw fracture and periodontitis are also responsible for oral infection.

Types

- **Primary tuberculosis**—it occurs in previously unexposed person and it involve lung.
- **Secondary tuberculosis**—in this reactivation of bacteria and it occur in, compromised host defense.
- **Miliary tuberculosis**—it spreads through bloodstream and there is wide involvement of many organs like kidney, liver and is called as miliary tuberculosis.
- **Pott's disease**—if tubercular involvement of spine occurs in children, it is called as Pott's disease.
- **Scrofula**—if it spreads by lymphatics to lymph nodes, it is called as scrofula.

Clinical Features

- **Age**—they are relatively uncommon and seen in middle and older age groups, as cleansing action of saliva and its antibacterial properties, in general also provide protection against tubercle bacilli.
- **Symptoms**—patient may suffer episodes of fever and chills, easy fatigability and malaise. There may be gradual loss of weight accompanied by persistent cough with or without hemoptysis. Local symptoms depend upon the tissue or organs involved.
- **Signs**—tubercular lymphadenitis may progress to acute abscess or remain as granulomatous lesion (**Fig. 30-8**). In any case, swelling of neck is present which is tender, painful and often show inflammation of the overlying skin. When abscess forms, it perforates and discharges pus.



Fig. 30-8: Tubercular lymphadenitis showing swelling in the submandibular region (Courtesy Dr Chole).

- *Pulmonary tuberculosis*—a persistence cough, hemoptysis, abundant sputum are usual features of pulmonary tuberculosis. There is also evening rise in temperature of 0.5° to 2°F. Night sweats is also present.
- *Scrofula*—in glandular form of the disease, there is marked enlargement of the cervical lymph nodes with caseation and frequent breakdown of the gland. Such tuberculous infection is called as 'scrofula'. It is usually caused by ingestion of unpasteurized infected cow milk.
- *Cold abscess*—the chronicity of the infection and the lack of marked pain or acute inflammatory symptoms have resulted in the term 'cold abscess'.
- *Consumption*—progressive tuberculosis may lead to wasting syndrome called as *consumption* as it appear as patient body is consumed or destroyed.
- *Lupus vulgaris*—involvement of skin in tuberculosis is term as lupus vulgaris.

Oral Manifestations

- *Site*—tongue is most commonly affected followed by palate, lips, buccal mucosa and gingiva.
- *Primary oral lesion*—it develops when bacteria is directly inoculated in the oral tissue of a person who has not acquired immunity to the disease. It involves gingiva, tooth extraction socket and buccal fold.
- *Secondary oral lesion*—infection is carried by hematogenous route or through break in the tissue surface, is deposited in the submucosa, subsequently proliferates and ulcerates the overlying mucosa. It occurs more frequently in cases of extra-pulmonary tuberculosis.
- *Tubercular ulcer*—the lesion may be preceded by an opalescent vesicle or nodule, a result of caseation necrosis. It breaks down into an ulcer which is usually superficial or deep and painful. Margin of the ulcer are undermined with minimum induration. It tends to be increased slowly in size. Mucosa surrounding the ulcer is inflamed and edematous. Base of ulcer is yellowish and granular.
- *Sentinel tubercle*—tiny, single and multiple nodules called 'sentinel tubercle' may also be seen surrounding the ulcer.
- *Mucocutaneous junction*—at the mucocutaneous junction, tubercular ulcers are usually extremely shallow with granulating base. Crusting and oozing is seen when the lesion involves adjacent cutaneous surface. They are usually painful.
- *Nodular form*—the nodular form of tuberculosis presents as single or multiple nodules of variable sizes, which initially may appear as semitransparent lesion of pinhead size. They are gray in size and of variable consistency.
- *Miliary tubercle*—miliary tubercle of oral mucosa are occasionally seen in miliary tuberculosis which is a result of acute dissemination of the infection through hematogenous and lymphatic channel. The oral lesion appears as small, gray tubercle with the tendency to break down and ulcerate.
- *Periapical tissue*—tubercle involvement of the periapical tissue and tooth socket have been reported. The socket may be filled with so called tuberculosis granulation tissue, consisting of many small, pink and red elevations.
- *Tuberculosis gingivitis*—tuberculosis gingivitis is an unusual form which may appear as diffuse, hyperemic or nodular papillary proliferation.
- *Jaw involvement*—it may involve maxilla or mandible. Jaw bone involvement occur as a result of either deep extension of a gingival lesion, from infected extraction socket, an extension of tubercular granuloma at the apex of tooth or by means of hematological spread. It may result in more diffuse form of osteomyelitis and is normally more serous than infections from periapical lesion.
- *Symptoms of jaw involvement*—tubercular involvement of jaw bone causes swelling and the symptoms include difficulty in eating, trismus, paresthesia of the lower lip and enlargement of the regional lymph nodes. Fistulae drain either intraorally or extraorally often with blue margins.
- *Temporomandibular joint*—in some cases, destructive involvement may reach to the TMJ area.

Radiographic Features

General

- *Early changes*—the chest radiograph in patient with secondary tuberculosis may show fibronodular changes most often in upper lobe. There is also cavity formation and volume loss. The earliest changes usually are ill defined opacity or opacities situated in the upper lobes.
- *Advanced cases*—in more advanced cases, opacities are larger and more widespread and may be bilateral. An area of translucency within the opacities indicates cavitations.
- *Displacement of trachea*—trachea and mediastinal structures are displaced towards the side of the lesion.

Oral

- It may be described as a localized or diffused with wide variation in size.
- *Location*—mandibles show greater predisposition than maxilla.
- *Localized lesion*—the localized lesion is of rarefying osteitis, while diffuse one is the nature of osteomyelitis.
- *Diffuse lesion*—in diffuse type of tuberculosis of bone, scattered area of bone destruction separated by portions of bone having normal or near normal appearance are seen.

- *Periostitis*—in some cases, tuberculosis may result in laying down the bone which resembles periostitis.

Diagnosis

- *Microscopy*—a pulmonary TB suspect, should submit 3 sputum samples for microscopy. Morning sample is ideal.
- *Staining method*—Ziehl-Neelsen, carbol fuschin or kinjoun carbol fuschin have been used for staining mycobacterium.
- *Polymerase chain reaction*—this technique amplifies even very small proteins of predetermined target region of *Mycobacterium tuberculosis* complex DNA.
- *Tuberculin skin test*—the Mantoux test is the preferred skin test for detecting tuberculosis. It involves the injection of 5 Tuberculin unit of purified protein derivatives, usually 0.1 ml intradermal. The skin test is read on the basis of millimeters of induration produced by PPD. 10-15 mm induration is required for the test to be positive.

Dental Considerations

- *Evaluation of patient*—Dental personnels are at constant risk of contacting the disease while treating the patient of tuberculosis. It is known that smallest droplet of contaminated saliva from the patient may be sufficient source of contagion, particularly if they are inhaled by previously uninfected or non-immunized individual. Patient with past history of tuberculosis should be evaluated by the physician to ensure the course of the treatment and follow up.
- *Delayed dental treatment*—if the culture is positive, dental treatment should not be performed unless there is emergency. In emergency dental treatment special precautions should be taken.
- *Protection of doctors*—to avoid aerosolization use of rubber dam and minimum use of ultrasonic scalers and high speed handpieces should be done. The operating air should be vented to the outside, i.e. not recirculated.

Differential Diagnosis

- *Syphilis*—primary lesion is harder and spirochetes are found.
- *Traumatic ulcer*—short duration and history of trauma.
- *Sarcoidosis*—tuberculin test negative, the granuloma of sarcoidosis are of non-necrotizing type.
- *Lupus erythematosus*—histology, causative agent test negative, skin changes.
- *Leukemia*—immature WBCs, prolonged bleeding and clotting time.

Management

- *Chemotherapy*—short term chemotherapy, isoniazid (5 mg/kg with maximum of 300 mg daily or 15 mg/kg two to three times weekly) and rifampicin (10 gm/kg), ethambutol (25 gm/kg daily for not more than 2 months).
- *Other drugs*—other drugs which can be used are streptomycin, para-aminosalicylic acid, pyrazinamide, thiacetazone, ethionamide and cycloserine.
- *Surgery*—surgery is rarely performed nowadays, as most patients respond well to medication. When indicated, it is usually for a persisting pulmonary cavity with acid-fast positive sputum despite 6 months of chemotherapy. The usual type of surgery done is segmental resection of lung or a complete lobectomy.

Actinomycosis

It is a chronic granulomatous suppurative and fibrous type of disease caused by anaerobic, gram +ve, non-acid fast bacteria. Most common are *Actinomyces israelii*, *A. nasalandi*, *A. viscosus* and *A. odontolyticus*. The organism is considered to be transitional form between bacteria and fungi. The term Actinomyces was given by Harz to refer the 'ray-like appearance' of the organism in the granule.

Classification

- *Cervicofacial*—it involve facial and cervical region.
- *Abdominal*—it is serious form and involves abdomen.
- *Pulmonary*—in this, there is pleural invasion resulting in empyema.
- *Cutaneous*—there is subcutaneous swelling in this form.
- *Central*—here, the infection is from the tooth or its membrane and is accompanied by radiographic changes.
- *Peripheral*—the peripheral types originate in the soft tissues and do not involve bone.

Predisposing Factors

- *Trauma*—the breach in the continuity of mucosa caused either by trauma or surgery, if the prerequisite for majority of actinomycosis infections.
- *Local factors*—cervicofacial actinomycosis infections are endogenous in origin and occur when dental plaque, calculus or gingival debris contaminate the relatively deep wounds around the mouth. Presence of carious teeth may act as predisposing factors for actinomycosis.
- *Others*—secondary bacterial invasion and hypersensitivity reaction may act as predisposing factors for actinomycosis.

Clinical Features

Cervicofacial form

- **Age**—it is most common type of actinomycosis and is commonly seen in adult males.
- **Location**—submandibular region is the most frequent site of infection. It usually spreads by direct tissue extension. Cheek and masseter region and parotid gland may also be involved.
- **Symptoms**—trismus is a common feature, before the formation of pus.
- **Signs**—the first sign of infection is characterized by the presence of a palpable mass. Mass is painless and indurated.
- **Sinus and fistula**—development of fistula and sinus is common. Skin surrounding the fistula is purplish (**Fig. 30-9**). Adjacent tissues have doughy consistency.
- **Sulfur granules**—several hard circumscribed tumors like swelling may develop and undergo breakdown, discharging a yellow fluid containing the characteristic submicroscopic sulfur granules.
- **Ray fungus appearance**—round or lobulated colony mesh-work of filaments stain with hematoxylin and peripheral club shaped ends of filaments stain with eosin.

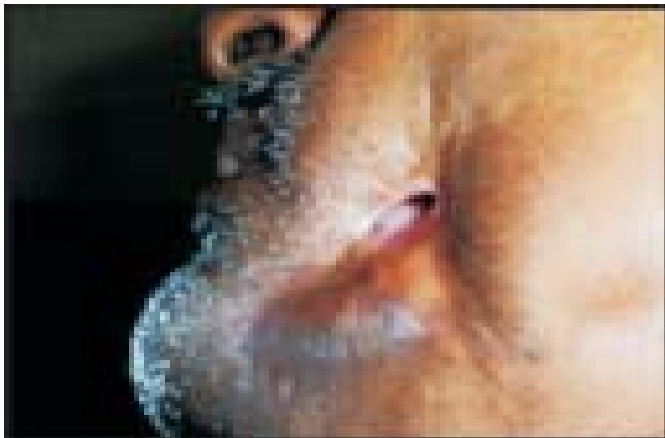


Fig. 30-9: Fistula seen in patient of actinomycosis (Courtesy Dr Patle).

Abdominal form

- Abdominal actinomycosis is extremely serious form of the disease.
- **Precipitating factors**—trauma, due to surgery or other causes such as fish bone or chicken bone injury, usually precedes the onset of the disease.
- **Symptoms**—generalized symptoms of fever, chills, vomiting, develop followed by symptoms of involvement of organs, such as liver and spleen.
- **Sign**—there is palpable abdominal mass. Intestinal manifestations develop later.

Pulmonary form

- **Symptoms**—it produces findings such as fever and chills, accompanied by a productive cough and pleural pain.

- **Signs**—pleural invasion resulting in empyema and there may be formation of sinus.

Subcutaneous form

- **Cause**—infection result from traumatic transplantation of organism, usually due to human bites.
- **Signs and symptoms**—lesion seen as subcutaneous swelling which enlarges slowly softens and ruptures through the sinuses. Occasionally, these lesions burrow through deeper tissue and invade bones.

Oral Manifestations

- **Cause**—organism may enter the tissue through oral mucous membrane and may either remain localized in the adjacent soft tissue or spread to involve salivary glands, bone or skin of face and neck.
- **Signs**—it produces swelling and induration of tissue. It may develop into one or more abscesses, which tend to discharge upon the skin surface liberating pus, which contains typical sulfur granules. There may be non-healing tooth socket, exuberant granulation tissue and periosteal thickening of alveolus. Skin overlying abscess is purple red and indurate or fluctuant.
- **Sinus**—it is common for sinus, through which the abscess has drained, to heal but due to chronicity, new abscesses are formed and perforate through skin surface.
- **Face**—there is disfigurement of face. Infection may involve maxilla and mandible.
- **Periapical granuloma**—there is formation of periapical granuloma.
- **Tongue**—on the tongue, the lesion is a painful nodule which eventually ulcerates. Untreated cases may reach to the point where the tongue may become fixed.
- **Actinomycosis osteomyelitis**—it can occur in patient with periodontal infection, nonvital teeth.

Radiographic Features

- **Appearance**—it may appear as an area of bone destruction, which resembles a dental cyst, with a well defined area of radiolucency with cortical lining of dense bone.
- **Lamina dura**—lamina dura is deficient at the apex of tooth.
- **Rarefying osteitis**—scattered area of bone destruction, separated from one another by normal or sclerosed bone, is another manifestation.
- **Tooth features**—the persistence radiolucency of tooth socket with an increased density of adjacent bone may be the first sign of disease.

Diagnosis

- **Clinical dagnosis**—sulphur granules with development of fistula and sinus with fever give clue to diagnosis.

- *Radiological diagnosis*—loss of lumina dura with scattered area of bone destruction.
- *Laboratory diagnosis*—typical key fungus appearance is seen.

Management

- *Two fold therapy*—actinomyces infection produces a reactive inflammatory response which causes an area of necrosis and scar tissue around the abscess. This results in decrease in the vascular supply to the affected region and hence, makes penetration of antibiotics difficult. Therefore, two fold therapy including antibiotics and surgery is necessary.
- *Surgical*—the lesion should be surgically removed and the surrounding area should be thoroughly debride.
- *Antibiotics*—following surgical intervention, antibiotics therapy should be continued. The antibiotic of choice is penicillin which should be given 3 to 4 million IV, every 4 hours for 2 to 4 weeks. This should be followed by 0.5 to 12 gm of penicillin, four times a day for 4 to 6 weeks. In patients allergic to penicillin, tetracycline orally 500 mg given four times a day for 6 months.
- *Other drugs*—other agents used in the treatment of actinomycosis include cephalosporin, clindamycin and lincomycin.

Noma

It is also called as 'Cancrum oris', 'gangrenous stomatitis', 'necrotizing stomatitis'. Noma is derived from the word *nomein* meaning to devour (eat greedily). It is rapidly spreading gangrene of oral and facial tissues occurring usually in debilitated or nutritionally deficient person. It is caused by *Fusobacterium necrophorum*.

Predisposing Factors

- *Poverty*—it occur in persons who are undernourished.
- *Debilitated disease*—debilitated infections such as diphtheria, dysentery, measles, pneumonia, scarlet fever, syphilis, measles, tuberculosis and blood dyscrasias.
- *Injury*—excessive mechanical injury is also predisposing factors.
- *Poor oral hygiene*—this may lead to growth of the bacteria causing increased susceptibility for the infection.
- *AIDS*—as it is immunological disorders, it can lead to Noma.
- *Others*—miscellaneous factors such as leukemia, sickle cell trait, stress and chemotherapeutic agents can cause noma.

Clinical Features

- *Age*—it is seen chiefly in children, but can be found in adults in certain conditions like in malnourished states.

- *Sites*—common sites are areas of stagnation around fixed bridge or crown.
- *Onset*—the commencement of gangrene is denoted by blackening of skin.
- *Necrotizing ulcerative mucositis*—small ulcers of gingival mucosa spread rapidly and involves the surrounding tissues of jaws, lips and cheeks by gangrenous necrosis.
- *Symptoms*—lesion has foul odor. Patients have high temperature during the course of the disease, suffer secondary infection and may die from toxemia or pneumonia.
- *Skin*—overlying skin is inflamed, edematous and finely necrotic which results in formation of line of demarcation between healthy and dead tissue.
- *Advanced stage*—in advanced stage, there is blue-black discoloration of the skin. As gangrenous process advances, slough appears and soon separated, leaving a perforating wound in the involved area (**Fig. 30-10**).
- *Noma neonatorum*—it arise in first month of low birth weight infants who are having malnutrition. Infants have lesion on lips, nose, mouth and anal area.
- *Jaw*—large masses may be sloughed out leaving the jaws exposed.
- *Complication*—it includes pneumonia, diarrhea, and septicemia. This can lead to death of patient.

Diagnosis

- *Clinical diagnosis*—blue black discoloration of skin with sloughing of tissue will give clue to diagnosis.



Fig. 30-10: Blackish discoloration with ulceration seen in case of gangrenous stomatitis (Courtesy Dr Tapasya).

Management

- *Control of dehydration and electrolyte balance*—parenteral fluid should be given urgently to correct dehydration and electrolyte balance.
- *Blood transfusion*—blood transfusion helps in improving the clinical state of the patient, who is usually anemic and toxic.
- *Antibiotics*—the specific drug of choice is penicillin although sulphonamides also yield good results. Other antibiotics which are used are gentamicin, clindamycin.
- *Reconstructive surgery*—reconstructive surgery is necessary to lead near normal life.

Scarlet Fever

Predominately occurs in children during winter months, caused by infection with group-A streptococci of beta hemolytic type that elaborate erythrogenic toxins.

Clinical Features

- *Incubation period*—incubation period is 1 to 7 days.
- *Symptoms*—patient exhibits severe pharyngitis and tonsillitis, chills, fever and vomiting.
- *Signs*—throat becomes highly erythematous and exudation is common. There may be enlargement and tenderness of regional lymph nodes.
- *Skin rash*—characteristic diffuse, bright scarlet to dusky red skin rash which appear on the second or third day of the illness (**Fig. 30-11**). This rash is also called as 'sunburn with goose pimples'. Rash is more intense in areas of pressure and skin fold. There is a transverse red streak which is called as 'Pastia's line'. After 3 to 4 days, the rash fades. This rash is due to toxic injury to the vascular endothelium which produces dilation of the small blood vessels and consequent hyperemia.



Fig. 30-11: Rash of scarlet fever seen on the skin of the hand (Courtesy Dr Pincha).

Oral Manifestations

- *Stomatitis scarlatina* accounts for the chief oral manifestation of scarlet fever.
- *Palate*—mucosa of palate may appear congested. Ulceration may also occur on palate.
- *Face*—the face, especially the temporal region and cheeks are flushed and red, but pale area of circumoral pallor is often seen around the mouth.
- *White strawberry tongue*—tongue exhibits white coating and fungiform papillae are edematous and hyperemic, projecting above the surface as small red knobs and it is called as 'strawberry tongue'.
- *Red raspberry tongue*—coating of tongue is soon lost, beginning at the tip and lateral margins and the organ becomes deep red, glistening and smooth, except for swollen hyperemic papilla. The tongue in this phase is called as 'raspberry tongue'.
- *Teeth*—in some cases, hypoplasia of teeth is seen in permanent teeth, if conditions occur at the time of tooth development.

Complications

- *Hypersensitivity reaction*—localized or generalized bacterial dissemination or hypersensitivity reaction to the bacterial toxins.
- *Systemic complication*—peritonsillar abscess, rhinitis, sinusitis, otitis media, mastoiditis, meningitis, pneumonia, glomerulonephritis, rheumatic fever and arthritis.
- *Oral complication*—it includes cancrum oris, ulceration with perforation of palate, osteomyelitis and involvement of the temporomandibular joint.

Diagnosis

- *Clinical diagnosis*—white strawberry and red raspberry tongue are typical of scarlet fever.

Differential Diagnosis

- *Pernicious anemia*—tongue in pernicious anemia is not as red as in scarlet fever.
- *Atrophic glossitis in vitamin deficiency*—swab culture of oropharynx disclose the presence of hemolytic streptococci and antistreptolysin -O titer is elevated.

Management

- *Antibiotics*—penicillin is the drug of choice, since group A streptococci are generally highly sensitive to this antibiotic. The species are also sensitive to other antibiotics like erythromycin, tetracycline and chloramphenicol.

Diphtheria

It is an acute contagious disease caused by gram +ve bacillus *Corynebacterium diphtheriae*, also called as *Klebs*

Loeffler bacillus. It is transmitted by droplet infection or direct contact.

Pathogenesis

- *Portal of entry*—the portal of entry is the upper respiratory tract and rarely skin, genitalia, eye and middle ear.
- *Incubation of bacteria*—the bacilli settle on the mucous membrane or upper respiratory tract and lead to inflammation and necrosis of mucosal cells. The infection may spread to adjacent areas.
- *Primary area*—in the primary invasive region, it forms a thick, firm, leathery, blue white pseudomembrane composed of bacteria, necrotic epithelium, macrophages and fibrin. A narrow zone of inflammation surrounding the area is seen.
- *Secondary area*—when the diphtheria bacteria multiply in the local tissues, they produce powerful exotoxins. This exotoxin diffuses through the body through a hematogenous route. Heart, muscle, kidney, peripheral nerves and adrenal glands are thus involved. Death may be caused by heart failure, airway obstruction which is caused by edema or by the effect of toxin.

Clinical Features

- *Age*—it occurs most frequently in children, during the fall and winter months.
- *Incubation periods*—incubation period is two days.
- *Symptoms*—listlessness, malaise, headache, fever and occasional vomiting. Within a short time, patient complains of sore throat.
- *Signs*—mild redness and edema of pharynx with cervical lymphadenopathy.
- *Bull neck*—there may be swelling of the neck, called as 'bull neck'.
- *Nose*—soft palatal paralysis can lead to nasal regurgitation of liquids during drinking.
- *Larynx*—larynx is edematous and is covered by pseudo-membrane. It produces a mechanical respiratory obstruction and typical croup.
- *Neural involvement*—there is generalized polyneuritis with weakness; paresthesia may follow in the next 10 to 14 days.
- *Complication*—it includes myocarditis, polyneuritis and acute intestinal nephritis.

Oral Manifestations

- *Diphtheritic membrane*—formation of patchy 'diphtheritic membrane' which begins on tonsils and enlarges, becoming confluent over the surface. It is thick and grayish in color.

- *Signs*—it tends to adhere and leave a raw bleeding surface on removal.
- *Soft palate*—soft palate temporary paralysis usually during 3rd and 5th week of the disease. The paralysis usually disappears in a few weeks or few months.

Diagnosis

- *Clinical diagnosis*—bull neck, sore throat with edematous larynx will give clue to diagnosis.

Prevention

- The disease may be prevented by prophylactic active immunization with diphtheria toxoid.

Differential Diagnosis

- *Herpes simplex infection*—small blisters, small shallow ulceration and history of prodromal symptoms. No patch on soft palate.
- *Hand-foot-mouth disease*—nausea, diarrhea, fever and vesiculoulcerative lesion occur simultaneously in the oral cavity and on hand and feet. No patch is seen.

Management

- *Rest*—the patient should be isolated and bed rest is very essential.
- *Antitoxin*—it is treated with diphtheria antitoxin. Mild cases treated with 10,000 to 20,000 units of antitoxin. Moderate cases with 20,000 to 40,000 units and severe cases with 50,000 to 100,000 units of antitoxin.
- *Antibiotics*—along with antitoxin antibiotics like penicillin and erythromycin should be given.

Cat Scratch Disease

It is also called as 'cat scratch fever'. This is most common cause of chronic regional lymphadenopathy. The etiological agent is unknown. Viral as well as bacterial agents have been proposed as the cause of this disease. Initially, causative organism is named as *Rochalimaea henselae* but later on, it is named as *Bartonella henselae*.

Clinical Features

- *Age*—it occur at any age, but predominant in children and young adults. It is thought to arise after traumatic break in skin due to scratch or bite of household cat.
- *Incubation period*—the incubation time of the disease ranges from 3-14 days.
- *Symptoms*—in early stage, low grade fever, headache, chills, nausea, malaise or even abdominal pain may occur.

- **Signs**—within few days of indolent primary lesion, often papules or vesicle develop at the site of injury.
- **Lymphadenitis**—within one to three weeks, lymphadenitis develops. The nodes are painful and may be several centimeter in diameter. The overlying skin may be inflamed. The lymph nodes gradually become soft and fluctuant, owing to necrosis and suppuration. The lymphadenopathy may persist for one to six months. In dental point of view, there may be involvement of preauricular, submaxillary or cervical chain of nodes.
- **Parotid pain**—when scratch occur in parotid area, infection may become localized parotid lymphoid tissue causing significant parotid pain.
- **Oculoglandular syndrome of Parinaud**—it consist of conjunctival granuloma with pre-auricular lymphadenopathy. It occurs when person touches fur moistened with cat saliva during cleaning. In this case, person touches the eye organism may get transfer into conjunctiva.
- **Others feature**—other manifestations includes non-pruritic macular or maculopapular rash, thrombocytopenia, pneumonia, conjunctivitis and grand mal seizure.

Diagnostic Criteria

Out of the following four, three should be positive for the diagnosis of cat scratch disease:

- Contact with cat with presence of scratch. Dermal and ocular lesion should be present.
- Negative results for other cause of lymphadenopathy.
- Characteristic histopathological finding of staining of pleomorphic bacilli.
- Positive cat scratch disease skin test (no longer widely used).

Management

- **Self limiting**—the disease has a benign course and it is self limiting.
- **Symptomatic treatment**—symptomatic treatment is given in the form of analgesic.
- **Antibiotics**—erythromycin is first choice of antibiotics. Another antibiotic which can be given is Doxycycline.
- **Node aspiration and drainage**—aspiration of the nodes should be done with needle tunneled laterally in the node.

Tularemia

It is also called as 'Rabbit fever'. It is caused by gram negative bacillus *Francisella tularensis*. It is contacted through infected rabbits, muskrats, ground squirrels and other wild germ, particularly of rodent family.

Types

- Cutaneous
- Ophthalmic
- Pleuropulmonary
- Oral
- Abdominal

Clinical Features

- **Incubation period**—incubation period is up to seven days.
- **Symptoms**—patient complaint of sudden headache, nausea, bony pain, profuse sweating, vomiting, chills and fever.
- **Suppurative ulcer**—a single cut or sore on the skin develops into a suppurative ulcer.
- **Lymph nodes**—lymphatic vessels become swollen and painful and the lymph nodes are remarkably enlarged.
- **Eyes**—the eyes also become involved with conjunctivitis developing through localization of disease in the conjunctival sac.
- **Complications**—tularemia pneumonia and pleuritis are complications of this disease.

Oral Manifestations

- **Cause**—primary infection of the mouth usually occurs from eating infected meat.
- **Site**—it is common on soft palate, tongue, gingiva and angle of mouth.
- **Symptoms**—severe pain is major complaint of the patient.
- **Diphtheria-like appearance**—the appearance of the ulcer is shallow with whitish fibrinous pseudomembrane formation. The tonsil, posterior pharyngeal wall, soft palate, base of the tongue and buccal mucosa may be covered by a grayish white membrane simulating the appearance of diphtheria.
- **Generalized stomatitis**—there may be generalized stomatitis.
- **Regional lymph nodes**—regional lymphadenitis may arise in submaxillary and the cervical groups of nodes. Cervical lymph nodes are tender, enlarged and may suppurate.

Diagnosis

- **Clinical diagnosis**—severe pain with ulcer with systemic symptom of headache, fever may give clue to the diagnosis.
- **Serology**—an agglutination test is used to demonstrate a rising titer of antibody in the serum of patients of *F. tularensis*.
- **Skin testing**—intradermal injection of an extract of *F. tularensis* gives a positive delayed hypersensitivity reaction in 1st or 2nd week of illness.

Management

- **Antibiotics**—antibiotics of choice are streptomycin and tetracycline, either alone or in combination.

Tetanus

It is also called as 'lock jaw'. It is a disease of nervous system characterized by intense activity of motor neurons and resulting in severe muscle spasm.

Pathogenesis

- **Organism responsible**—it is caused by anaerobic gram positive bacillus *Clostridium tetani*.
- **Production of toxins**—the spores of above organism germinated under the anaerobic condition. This will result in production of exotoxin, tetanospasmin and hemolysis.
- **Toxins reach the brain**—all of the above toxins reach the brainstem and anterior horn of spinal cord via bloodstream.
- **Increase muscle tone**—toxin increases muscle tone and rigidity resulting in muscle spasm and convulsion.

Transmission

- **Injury**—it can enter the body through the most trivial injury.
- **Intravenous drug user**—it is often reported amongst intravenous drug users.
- **Contamination**—it is transmitted through contaminated soil, dust or wood splinters.
- **Tetanus neonatorum**—infection of the cut surface of umbilical cord or circumcision wound, due to use of unclean instruments or dressing, may result in tetanus of the newborn which is called as 'tetanus neonatorum'.

Types

- **Local tetanus**—muscle spasm at the site of entry is known as local tetanus.
- **Generalized form**—this type of tetanus involve many system of the body.
- **Cephalic form**—this type of tetanus occurs in association with facial palsy.
- **Neonatal form**—this occurs in infant and route of transmission is from mother.
- **Chronic form**—the cause of it is due to the persistence of focus of infection and fibrosis from inadequately controlled spasms.

Clinical Features

- **Age and sex**—it is more common in young males during their accident prone years.

- **Incubation period**—the clinical manifestations occur within 14 days after incubation period.
- **Local tetanus**—it is characterized by muscle spasm near the site of entry of bacilli. In some cases, it may proceed to generalized form. Mortality rate is less than 1% in this form.
- **Cephalic form**—in this, bacilli are introduced through the wounds in the head and neck region. *Cranial nerve palsy* occurs with cranial nerve, most commonly the 7th cranial nerve.
- **Generalized form**—entire body may be affected with contraction of all groups of somatic muscles leading to characteristic *opisthotonos* (spine become arch shaped). Reflex spasm frequently develops after stimuli. The temperature is usually raised due to increase metabolic rate. When spasm of intercostal, pharyngeal and diaphragmatic muscle occurs, adequate ventilation becomes impossible and the resultant anoxia causes death. Death in patients with tetanus is generally related to pulmonary complications including broncho-pneumonia and pulmonary embolism.
- **Neonatal tetanus**—the incubation period of neonatal tetanus is 3-10 days. It has high fatality rate. The disease is characterized by difficulty in suckling and excessive crying in early stage. As the disease progresses, variable degree of muscular spasm develops. Death is usually due to inadequate ventilation and asphyxia.
- **Chronic tetanus**—it may persist for many years. Clinically, there are features of generalized tetanus to intermittent spasm of individual muscles.

Oral Manifestation

- **Rigidity of muscle of mastication**—this is usually first manifestation of tetanus.
- **Risus sardonicus**—rigidity of facial muscles may occur, producing the typical 'risus sardonicus'. In this, corners of mouth are drawn back with protruded lip, wrinkling of forehead not possible.
- **Symptoms**—patient complaint of difficulty in chewing, swallowing and inability to insert denture.
- **Lock jaw**—as spasm of muscle of mastication increase jaw is locked and mouth cannot be open.

Diagnosis

- **Clinical diagnosis**—lock jaw with risus sardonicus and opisthotonus will give clue to the diagnosis.

Management

- **Antitoxin**—neutralizing the toxin is achieved by administered of human tetanus immunoglobulin. Specific therapy includes immediate intravenous injection of immune serum containing 20,000 IU of antitoxin.

- **Penicillin**—1,000,000 units of penicillin G intravenously, every 6 hours, for 10 days must be administered to kill vegetative form of *Clostridium tetani*.
- **Sedation**—sedation of patient should be done with diazepam.
- **Supportive care**—supportive care includes providing proper nutrition.

Rhinoscleroma

It is unusual chronic infection caused by bacillus *Klebsiella rhinoscleromatis* and it is common in Europe and central and South America.

Clinical Features

- **Age and sex**—both sexes are equally affected. Common between the ages of 20 to 40 years.
- **Site**—usually found in upper respiratory tract, often originating from nose, but involvement of lacrimal glands, orbit, skin, paranasal sinus and intracranial invasion have been described.
- **Rhinitis stage**—in this stage, there is nasal obstruction, nasal deformity and epistaxis. There is also swelling of upper lip and sore throat.
- **Infiltrative stage**—it is characterized by nasal obstruction, due to the presence of exuberant granulation tissue. Hoarseness results due to laryngeal involvement. Anesthesia of soft palate is common in this stage.
- **Nodular stage**—the proliferation of nasal masses may produce configuration known as 'Hebra nose'. Posterior extension of the lesion may produce laryngeal and tracheal obstruction of varying degrees. Complications include scleromatous infiltration of eustachian tube and unilateral scleroma of maxillary sinus.

Oral Manifestations

- **Site**—it is common on lip, soft palate, and tonsil.
- **Appearance**—they appear as proliferative granulomatous lesion of lip, soft palate and tonsil.
- **Symptoms**—there is anesthesia of soft palate, impaired taste, upper lip and uvula enlargement.

Diagnosis

- **Clinical diagnosis**—hebra nose with rhinitis and enlargement of lip, and anesthesia of soft palate will give clue to the diagnosis.

Management

- **Antibiotics**—it should be given to control infection.

Granuloma Inguinale

It is also called as '*granuloma venereum*' and '*donovanosis*'. It is found in inguinal and anogenital region and is caused by *Calymmatobacterium granulomatis* a gram negative bacillus with prominent polar granules. It is chronic slowly progressing, mildly contagious disease.

Clinical Features

- **Age and sex**—it chiefly affects adult black of either sex, but may occur in any race.
- **Appearance**—lesion begins as a small papule that ulcerates, increases in size and eventually gives rise to velvety, beefy, granulating and spreading ulcerative lesion of inguinal and anogenital region.
- **Pseudo-bubo**—inguinal ulceration is commonly secondary to the genital lesion and arises initially as fluctuant swelling known as pseudo-bubo.
- **Metastasis**—metastatic spread to bone and soft subcutaneous tissue has been reported.

Oral Manifestations

- **Cause**—oral lesion appears to be the most common extra-genital form of granuloma inguinale. Oral lesions occur either as a result of autoinoculation through infected fingers or through oral coitus.
- **Site**—it is most commonly found on lips, buccal mucosa or palate or they may diffusely involve the mucosal surface
- **Lip**—lesions of lip are characterized by extensive superficial ulceration with well defined elevated, granulomatous margins.
- **Multiple sinus**—long standing lesion is associated with multiple extraoral sinuses.
- **Types**—may be of three types, i.e. ulcerative, exuberant and cicatricial.
 - **Ulcerative**—it is painful, extensive with punched out appearance. It has got smooth red base with tendency of bleeding from the lesion.
 - **Exuberant**—it appears as proliferative granular mass. The surface of the lesion is rough due to projection. It has got sharp border and lesion are multiple. The mucous membrane is inflamed and edematous.
 - **Cicatricial**—fibrous scar formation may become extensive in nature.

Diagnosis

- **Clinical diagnosis**—ulcerative lesion in the oral cavity which can be multiple and exuberant in variety, will give clue to the diagnosis.

- **Laboratory diagnosis**—Donovan bodies present in smears which are recognized as large, gram negative oval bacteria with intense bipolar staining (*safety pin appearance*). There is granulation tissue with infiltration of polymorphonuclear leukocytes and plasma cells.

Management

- **Antibiotics**—azithromycin is the drug of choice. It should give intermittently. Treatment should continue until healing is complete which usually takes 2-3 weeks.
- **Wound debridement**—in chronic cases, wound debridement followed by antibiotics therapy is helpful.
- **Plastic surgery**—plastic surgery is needed for correction of extensive scarring.

Lymphogranuloma Venereum

It is a venereal disease caused by one of the three strains of *Chlamydia trachomatis*.

Clinical Features

- **Incubation period**—incubation period is 10 days.
- **Symptoms**—there may be fever, chills, headache and malaise.
- **Signs**—it persists as firm, tender enlargement of inguinal lymph nodes. Nodes are tender and adherent to the underlying tissues.
- **Groove sign**—enlargement of lymph nodes both above and below the inguinal ligament, is characteristic feature called as 'groove sign'.
- **Overlying skin**—overlying skin becomes reddened and dusky and multiple purulent fistulae develop over enlarged glands.
- **Females**—in females, placenta is frequently involved. Marked scarring and local edema frequently develops secondary to suppurative lymphadenitis.

Oral Manifestations

- **Causes**—results due to orogenital contact or anti-inoculation.
- **Site**—tongue is the most common site.
- **Appearance**—lesion consists of small, slightly painful superficial ulceration with non-indurated borders which appear on lip.
- **Signs**—in long standing infection, there is zone of cicatricial refraction, dark red area with loss of superficial epithelium, or opaque lichenoid grayish papules.
- **Tongue enlargement**—tongue may become enlarged with areas of scarring and grooves on its dorsal surface.
- **Associated symptoms and signs**—dysphagia, red soft palate and small red granulomatous lesions accompanied by

regional lymphadenopathy are commonly associated symptoms.

- **Lymph nodes**—cervical lymphadenopathy is present. Skin covering the swollen nodes is violaceous and indurated usually with one or more draining sinuses.

Diagnosis

- **Clinical diagnosis**—groove signs with tongue enlargement with scarring will give clue to the diagnosis.
- **Laboratory diagnosis**—lymph nodes biopsy will show minute intracellular cocci.

Management

- **Antibiotic therapy**—it consists of macrolides, tetracycline, and fluoroquinolones.
- **Surgical management**—surgical treatment of bubos and secondary scarring.

Myiasis

It is referred to the invasion of living tissues by the larvae of certain species of flies. Myiasis, the term first introduced by Hope (1840), refers to the invasion of living tissues of humans or other mammals by the eggs or larvae (maggots) of *Diptera* (two winged) flies.

Predisposing Factors

- **Poor oral hygiene**—it has been associated with poor oral hygiene, such as in mouth-breathers, thumb-suckers.
- **Mentally retarded person**—it is common in mentally retarded persons.
- **Cerebral palsy and hemiplegia**—this person cannot maintain normal lip closure or normal level of oral care.

Pathogenesis

- **Organism responsible**—myiasis is caused by members of the *Diptera* (two winged fly) family that lay over 100 to 500 eggs at a time—on food, necrotic tissue, open wounds and unbroken skin and mucosa.
- **Life cycle of larva**—the eggs hatch to become maggots in less than 1 week and the life cycle is completed when the maggots turn into flies in about 2 weeks.
- **Obtaining the nutrition**—the larvae obtain their nutrition from the surrounding tissues and burrow deeper into the soft tissues by making tunnels separating the gingiva and mucoperiosteum from the bone. Necrotic tissue, present in advanced periodontal diseases will form a good substrate in which the fly can lay its eggs. The periodontal pockets provide the eggs a warm and moist environment for further development.

Types

- *Cutaneous or dermal myiasis*—it affects skin.
- *Myiasis of external orifices*—it includes nasal, oral, vaginal and anal myiasis.
- *Myiasis of internal organs*—it includes intestinal and urinary myiasis.

Clinical Features

- *Cutaneous myiasis*—it is characterized by papular or migratory lesions.
- *Symptoms*—papular lesions are itchy and occasionally painful.
- *Open lesions*—open lesion may produce serious discharge and larvae may be detected through the opening (**Fig. 30-12**).
- *Migratory lesions*—migratory lesions are superficial, red tunnel-like lesions, which form creeping eruptions.



Fig. 30-12: Discharge seen from open lesion in oral myiasis.

Oral Manifestation

- *Incidence*—oral myiasis is a rare condition.
- *Appearance*—it presents as an erythematous, edematous or granulomatous lesion.
- *Symptoms*—itching or pain may be present.
- *Signs*—these lesions may pulsate with movement of larvae. An opening is present from which larvae can come to surface of the lesion (**Figs 30.13A and B**).

Diagnosis

- *Clinical diagnosis*—it is easy to make clinical diagnosing by observing larvae in the lesion.

Management

- *Irrigation*—irrigation of the tissue with hydrogen peroxide will come larvae out the lesion (**Fig. 30-14**). Treating the defect with ether, mercuric chloride, or turpentine oil, this irritates the parasites forcing them to



Figs 30.13A and B: Larvae seen in lesion of oral cavity (Courtesy Dr Bhaskar Patle).



Fig. 30-14: Larvae seen in the plate which are removed after patient given hydrogen peroxide (Courtesy Dr Bhaskar Patle).

come out. Whitehead varnish pack, which contains ether, can be applied to the raw wound for protection during the healing phase.

- **Surgery**—surgical removal of larvae should also be done.
- **Intestinal myiasis**—intestinal myiasis may require purgation with sodium sulphate or antihelminthics.

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Definition

Viruses have been defined as submicroscopic entities which reproduce within the specific living cells. Virus may contain either ribonucleic acid (RNA) or deoxyribonucleic acid (DNA).

Classification

RNA Virus

Orthomyxovirus

- Influenza

Paramyxovirus

- Measles (rubeola)
- Mumps

Rhabdovirus

- Rabies
- Hemorrhagic fever

Arenavirus

- Lymphocytic choriomeningitis
- Lassa fever

Calicivirus

Coronavirus

- Upper respiratory infection

Bunyavirus

Picornavirus

- Poliomyelitis
- Coxsackie diseases
- Common cold
- Foot and mouth disease
- Encephalomyocarditis

Reovirus

Togavirus

- Rubella
- Yellow fever
- St. Louis encephalitis

Retrovirus

DNA Virus

Herpes virus

- Herpes simplex I (herpes stomatitis and herpes labialis) or HHV 1
- Herpes simplex II (genital lesion) or HHV 2
- Herpes zoster (chicken pox and shingles) or HHV 3
- Epstein-Barr virus (EBV) (infectious mononucleosis, hepatitis, oral hairy leukoplakia and nasopharyngeal carcinoma) or HHV 4
- Cytomegalovirus (infectious mononucleosis, hepatitis) or HHV 5
- Human herpes virus -6 (roseola infantum, otitis media, encephalitis) or
- Human herpes virus -7 (roseola infantum)
- Human herpes virus -8 (infectious mononucleosis, febrile exanthema, Kaposi's sarcoma)
- Simian herpes virus B (mucocutaneous lesions, encephalitis)

Pox virus

- Small pox
- Molluscum contagiosum

Adenovirus

- Pharyngoconjunctival fever
- Epidemic keratoconjunctivitis

Parvovirus

Iridovirus

Papovavirus

- Human warts or papillomas
- Tumorigenic virus in animals

Infections Caused by Viruses

Herpes Simplex Infection

HSV is ubiquitous virus. It is the most common viral disease affecting men. Initial infection may be acquired from eyes, skin and mucosa with infected secretion. After acquiring

the infection, all herpes viruses remain latent in host neural cells, thereby evading host immune response.

Epidemiology

- *HSV I*—this virus mainly affect the region above the waist.
- *HSV II*—in this case, infection occurs below the waist usually in the genital lesion. But nowadays due to increased oral sex, HSV II is found in the oral lesions. Both HSV I and II can be transmitted sexually.
- *Occurrence*—it occurs in early childhood. Preschool period is more prone due to frequent exchange of salivary and nasal secretions.

Pathogenesis Oral Herpes

- *Primary infection*—the virus enters the skin or mucous membrane and reproduces. During this stage, oral signs and symptoms, such as fever, may develop. The virus may cause asymptomatic infections. Asymptomatic infection occurs twice as often as symptomatic disease.
- *Latency*—from the infected site, the virus moves to nerves in the region. There the virus reproduces and becomes inactive.
- *Recurrence*—during certain stresses, emotional or physical, the virus may reactivate and cause new signs and symptoms.

Types

- *Herpetic genitalis*—it is caused by HSV-II and is common in uterine cervix, vagina, vulva and penis. Lesions are more likely to occur on well keratinized areas such as shaft of penis but may progress, more or less rapidly, into superficial erosions that are very painful when touched or contaminated with urine. Urethra may be involved and extremely painful dysuria may develop, especially in women. Regional lymphadenopathy, fever, malaise and anorexia may develop.
- *Genital herpes in HIV patients*—the lesion of genital herpes in immunocompromised patients and HIV infected patients are larger, deeper and are likely to persist and release of virus for periods as long as 30 days. Antibodies to the virus develop 4 to 6 weeks after primary infection.
- *Herpetic meningoencephalitis*—this is serious form characterized by sudden fever and symptoms of increased intracranial pressure. Paralysis of various muscle groups with convulsion and even death may occur.
- *Herpetic conjunctivitis*—swelling and congestion of palpebral conjunctiva, keratitis and corneal ulceration. Recurrent lesions can lead to serious corneal scarring, which may produce blindness.
- *Herpetic eczema* (Kaposi varicelliform eruption)—it is epidermal form of herpetic infections superimposed

upon a pre-existing eczema and is characterized by diffuse vesicular lesions of skin. The patient usually exhibits a high fever coincident with typical umblicated vesicles as well as other systemic manifestation.

- *Disseminated herpes simplex infection of newborn*—it is uncommon and they acquired it during passage through the birth canal of mother suffering from herpetic vulvovaginitis. The newborns exhibit a wide variety of signs and symptoms of the disease and with few exceptions; die on the ninth or twelfth day of life.
- *Herpetic whitlow*—this is the infection of finger by herpes virus through the break in the skin. Dentist may experience primary lesion of fingers from contact with lesions of the mouth or saliva of the patients who are asymptomatic carriers of HSV; called as 'herpetic whitlow'.
- *Herpes gladiatorum or Scrumpox*—infection of skin spreads through the sport of wrestling.
- *Oral infection*—in the oral cavity, there may be *primary infection* (person who is without circulating antibodies) and *recurrent infection* (circulating antibodies are already present).

Primary Herpes Simplex Infection

It is also called as 'acute herpetic gingivo-stomatitis', 'herpes labialis', 'fever blister', 'cold sore' and 'infectious stomatitis'. It occurs in patients with no prior infection with HSV-1. HSV reaches nerve ganglion supplying the affected area, presumably along nerve pathways and remains latent until reactivated. The usual ganglion involved is the trigeminal for HSV-1 and lumbosacral, for HSV-2.

Transmission

- *Close contact*—it occurs during close personal contact, in which exchange of saliva or other secretion happened.
- *Newborn infection*—primary infection of newborn is believed to be caused by vaginal secretions during birth, which results in viremia and disseminated infection of brain, liver, adrenals and lungs.
- *Socioeconomic status*—incidence varies according to socioeconomic group. Person who is having lower economic status will have earlier exposure.

Clinical Features

- *Age*—it develops in both, children and young adults. It can be seen in high school and college students where it is transmitted by kissing and sexual contact.
- *Incubation period*—incubation period is 5 to 7 days.
- *Prodromal symptoms*—prodromal symptoms precede local lesion by 1 to 2 days and it includes fever, headache, malaise, nausea, vomiting and within a few days, mouth

becomes painful. There is also irritability, pain upon swallowing and regional lymphadenopathy.

- **Location**—this lesion mainly occurs on the hard palate, attached gingiva and dorsum of tongue. In some cases, lesion may be presented on the skin (**Fig. 31-1**).



Fig. 31-1: Showing skin lesion of herpes infection (Courtesy Dr Sanjay Pincha).

- **Appearance**—after this, small vesicles, which are thin walled, surrounded by inflammatory base are formed (**Fig. 31-2**). They quickly rupture leaving small, shallow, oval shaped discrete ulcers.
- **Size**—the individual ulcer differs in size from 2-6 mm. As the disease progresses, several lesions may coalesce, forming larger, irregular lesions.
- **Base**—the base of the ulcer is covered with grayish white or yellow plaque.
- **Margins**—the margins of the sloughed lesions are uneven and are accentuated by bright red rimmed, well demarcated, inflammatory halos.



Fig. 31-2: Intact vesicle seen in herpes simplex infection which is thin walled (Courtesy Dr Chole).

- **Lips**—in severe cases, excoriation involving the lips (**Fig. 31-3**) may become hemorrhagic (**Fig. 31-4**) and matted with serosanguinous fibrin-like exudate and parting of the lips during mastication and speech may become extremely painful and difficult.



Fig. 31-3: Lip lesion seen along with marginal gingivitis (Courtesy Dr Chole).



Fig. 31-4: Excoriation of lip with hemorrhagic lesion seen in herpes infection.

- **Acute marginal gingivitis**—appearance of generalized marginal acute gingivitis is typical feature of herpes simplex infection. Entire gingiva is edematous and swollen and small gingival ulcers are seen (**Fig. 31-5**).
- **Pharynx**—examination of posterior pharynx reveals inflammation causing difficulty in swallowing.
- **Lymph nodes**—cervical and submandibular lymphadenopathy is present.



Fig. 31-5: Acute marginal gingivitis present in the case of herpes simplex infection (Courtesy Dr Chole).

- **Healing**—the disease is self limiting and lesions begin healing in a week to 10 days and leave no scar.
- **Herpes simplex infection of adults**—it is seen in patients who do not acquire HSV infection and immunity during childhood. In such cases, primary herpes simplex infection is often acquired via new sexual partner and primary HSV gingivostomatitis and pharyngitis can occur. It occurs usually due to HSV I (Figs 31-6A and B)
- **Histopathological finding**—Lipschutz bodies can be seen in HSV infection. In cytologic study, there is presence of multinucleated giant cells, and ballooning degeneration of cells.

Recurrent or Secondary Herpetic Infection

In some cases, HSV may remain latent in epithelium which is the main cause of recurrent infection with HSV. In this case, HSV reactivated at the latent site it moves centrally to the mucosa. When reactivation is triggered, they spread along the nerves to different sites on oral mucosa and skin, destroy the epithelial cells and induce the typical inflammatory response with characteristic lesions of recurrent infection.

Types

- **Recurrent herpes labialis (RHL)**—If it occurs on lip, it is called as recurrent herpes labialis.
- **Recurrent intraoral herpes simplex infection (RIH)**—If occurs intraorally it is called as recurrent intraoral herpes infection.

Precipitating Factors

- **Surgery**—when any surgery which involve trigeminal ganglion, recurrent infection with herpes can occur. The



A



B

Figs 31-6A and B: Primary infection occurring in adult patient. (A) Ulcer seen with acute marginal gingivitis and (B) Large ulcer seen on the tongue.

reason for this is herpes virus remain latent in trigeminal ganglion.

- **Immunity**—low serum IgA, decreased cell mediated immunity, decreased anti-herpes activity and depression of ADCC (antibody dependent cellular cytotoxicity) and interleukin-2, caused by prostaglandin release in skin can precipitate the attacks.
- **Trauma**—trauma to lips, dental extraction can lead to recurrent infection.
- **Infection**—in some cases, upper respiratory tract infection can trigger the herpes infection.
- **Others**—fever, emotional upset, sunburns, fatigue, menstruation, pregnancy and allergy can be some precipitating factors for herpes infection.

Clinical Features

- **Occurrence**—recurrent herpes simplex infection may occur at widely varying intervals, from nearly every

month in some patients to only about once a year or even less in others.

- *Prodromal symptoms*—in either location, lesion is preceded by tingling and burning sensation and feeling of tautness, swelling or slight soreness subsequent development of vesicle.
- *Signs*—it is accompanied by edema at the site of the lesion, followed by formation of clusters of small vesicles.
- *Recurrent herpes labialis*—these gray or white vesicles rupture quickly leaving small red ulcerations, sometimes with slightly erythematous halo on lip covered by brownish crust on lips (**Fig. 31-7**). Sizes range from 1 to 3 mm in diameter, to 1 to 2 cm. But sometimes, it is large enough to cause disfigurement (**Fig. 31-8**).



Fig. 31-7: Recurrent herpes labialis showing vesicle of the lip (Courtesy Dr Amit Parate).



Fig. 31-8: Extensive lesion of herpes labialis showing disfigurement (Courtesy Dr Amit Parate).

- *Recurrent intraoral herpes*—in recurrent intraoral herpes (RIH) vesicles break rapidly to form small red ulceration, sometimes with slight erythematous halo. Cluster of

small vesicles or ulcers 1 to 2 mm in diameter are commonly found on gingivae, tongue (**Fig. 31-9**) palate and alveolar region.



Fig. 31-9: Recurrent intraoral, herpes infection seen on the tongue (Courtesy Dr Amit Parate).

- *Healing*—the lesions gradually heal within 7-10 days and leave no scars.
- *Complication*—herpes simplex infection can lead to development of extra-genital lesions, CNS complications and vaginal fungal super infections.

HSV in Immunocompromised Patient

It is described in *Chapter 35: AIDS*.

Diagnosis

- *History*—negative past history of recurrent herpes labialis and a positive history of close contact with a patient with primary or recurrent herpes is helpful in making the diagnosis.
- *Typical clinical features*—patient is easily diagnosed as having primary herpetic gingivostomatitis; if he/she presents with typical clinical features of generalized symptoms followed by eruption of oral vesicles and acute marginal gingivitis and does not have history of recurrent herpes.
- *HSV isolation*—isolation and neutralization of virus in tissue culture is most positive method of identification. Rabbit kidney and human amnion are sensitive to HSV.
- *Antibody titer*—antibodies to HSV appear in a week and react peak in 3 weeks.

Differential Diagnosis

- *Hand-foot and mouth disease*—in this case, lesions are not clustered and gingiva is not affected. Lesions can also be seen on feet and hand.

- *Herpangina*—oropharyngeal and soft palate involvement is more prominent. It affects children in late summer and early monsoon season on soft palate and facial area with fever and malaise.
- *Chronic recurring aphthae*—no stomatitis, no general systemic symptoms and lesions are less numerous and more often found in adults.
- *Herpes zoster*—segmental distribution along the anatomical location of nerve.
- *Erythema multiforme*—it occurs in young adults, as compared to herpes simplex which occurs in children. Gingivitis is not severe and generally limited to anterior part of the mouth. In erythema multiforme skin lesions are present.
- *Bullous lichen planus*—it is painful condition characterized by large blister on tongue and cheek with rupture and it undergo ulceration.
- *Cheilitis granulomatous*—edema appears suddenly without prodromal symptoms and recurrence is absent.
- *Benign mucosal pemphigoid*—no prodromal symptoms; with progression from erythema to swelling to blister to crusting.
- *Allergic contact dermatitis*—sudden appearance with allergen proof.

Management

Primary herpes simplex infection

Symptomatic

- *Pain control measures*—topical anesthetics like 2% lidocaine, 0.1% diclonine hydrochloride, 0.5% benzocaine hydrochloride are used. Solution of diphenylhydramine hydrochloride (*Benadryl*) 5 mg mixed with equal amount of milk of magnesia can also reduce the pain. In some cases systemic administration of analgesics is also given.
- *Topical anti-infective agents*—it is given to prevent secondary infection. Agents used are 0.2% chlorhexidine gluconate, tetracycline mouth wash and elixir or diphenylhydramine.
- *Supportive care*—fluid is given to maintain proper hydration and electrolyte balance. Antipyretics can also be given to control the fever.
- *Good oral hygiene*—oral hygiene should be properly maintained to avoid any secondary infection.

Specific

- *Acyclovir*—it inhibits DNA replication in HSV infected cells reducing the duration of illness but with few side effects. The optimum oral dosage of acyclovir is 1,000 to 1600 mg daily, for 7 to 10 days. It should be ideally given in a dose of 15 mg/kg five times a day.
- *Valacyclovir*—it is prodrug of acyclovir and it has far better biocompatibility as compared to acyclovir. It should be used in combination with famciclovir.

Recurrent herpes simplex infection

- *Minimized obvious trigger*—recurrent infection can be suppressed by reducing trigger factors. This can be done by applying the sunscreen lotion to lip.
- *Topical antiviral medication*—topical antiviral medication like 5% acyclovir cream, 3% penciclovir cream, 10% docosanol cream are effective. It should be applied three to six times daily.
- *Topical ammonium chlorides*—nowadays, recent formulation of quaternary ammonium chloride, dimethyl carbonal is used and has been marketed. But it is not scientifically proved to be effective so its use not recommended.
- *Systemic antiviral medication*—usually Valacyclovir or famciclovir (500-1000 mg three times daily) should be given.
- *Topical carbon oxolone*—it is useful in some cases of herpetic gingivostomatitis.

Varicella Zoster Infection

It is an acute disease caused by varicella zoster virus, which is a DNA virus similar to HSV and causes both, primary and recurrent infection. After primary disease is healed, VZV becomes latent in the dorsal root ganglion of spinal nerve or extramedullary ganglion of cranial nerve. VZV becomes reactivated causing lesions of localized herpes zoster. Patients with HIV infection, leukemia and those on immunosuppressive therapy have an increased susceptibility to severe or potentially fatal herpes zoster. Herpes zoster infection can be deep seated and disseminated causing pneumonia, meningoencephalitis and hepatitis.

Forms

- *Chickenpox (Varicella)*—it is primary infection with zoster virus.
- *Shingles (herpes zoster) or zona*—it is recurrent infection with zoster virus. It increases with age and occurs in immunocompromised patient. It can be seen in AIDS patient.
- *Postherpetic neuralgia*—it is neuropathic disease resulting from peripheral and central nervous injury by zoster virus.
- *James Ramsay Hunt syndrome*—it is zoster infection of geniculate ganglion.

Chickenpox

It is also called as 'varicella'. It is an acute viral disease occurring in children and most commonly in winter and spring months.

Clinical Features

- *Incubation period and mode of transmission*—incubation period is two weeks and mode of transmission is by air borne droplet or direct contact with infected persons, with the probable port of entry being respiratory tract.
- *Age*—it is seen in first two decade of life.
- *Prodromal symptoms*—it is characterized by prodromal occurrence of headache, nasopharyngitis and anorexia, followed by maculopapular or vesicular eruptions on skin and low grade fever.
- *Location*—these eruptions usually begin on the trunk and spread to involve the face and extremities.
- *Onset*—there is development of a pruritis, maculopapular rash followed by vesicle.
- *Dewdrop on rose petal*—there is classic presentation of centrally located vesicle surrounded by zone of erythema. This is called as *dewdrop on rose petal* appearance (Fig. 31-10). They occur in successive crops so many vesicles in different stages of formation or resorption may be found.
- *Healing*—the skin eventually ruptures, forming a superficial crust and heals by desquamation. The disease runs its clinical course in a week to ten days, seldom leaving any after effects.
- *Secondary infection*—occasionally, secondary infection of vesicle results in the formation of pustules which may leave small pitting scar upon healing.
- *Complication*—it includes cerebellar ataxia, encephalitis, pneumonia, myocarditis, Reye's syndrome, sickle cell crisis, hemolytic anemia and hepatitis.
- *Neonatal chickenpox*—this occurs due to infection in pregnancy. This infection may lead to congenital defect in the newborn.
- *Diagnosis*—it is made by virus isolation and typical exanthema present.



Fig. 31-10: Dewdrop like vesicle formation seen on the abdomen of the patient (Courtesy Dr Pincha).

Oral Manifestation

- *Site*—small blister-like lesions occasionally involve the oral mucosa chiefly buccal mucosa, tongue, gingiva, palate as well as the mucosa of pharynx.

- *Appearance*—the mucosal lesion, initially a slightly raised vesicle with a surrounding erythema, ruptures soon after formation and forms a small eroded ulcer with red margins (Fig. 31-11), closely resembling aphthous lesion.



Fig. 31-11: Intraoral ulceration seen in case of chickenpox infection (Courtesy Dr Sanjay Pincha).

Management

- *Pain control*—patient should be managed with all pain control method which is described in herpes simplex infection. Aspirin is contraindicated as it can lead to Reye's syndrome.
- *Control of pruritus*—warm baths with soap or baking soda, application of calamine lotion should be done. As VZV is having lipid envelope, it can destroy by soap and detergent. Systemic diphenhydramine is also given to control pruritus.
- *Antiviral drug*—acyclovir should be used in a dose of 800 mg five times daily. It will reduce the severity of lesion. Another antiviral agents use are valacyclovir (1000 mg TDS) and famciclovir (500 mg TDS) for 7 days should be given.
- *Varicella zoster immune globulin*—this is given in case of immunocompromised patient. This will help to reduce severity of clinical manifestation.
- *Local antiseptic*—at the first sign of secondary infection, a local antiseptic should be applied to the skin. If the bacterial infection progresses, appropriate antibiotics should be prescribed.
- *Prevention by vaccination*—live attenuated VZV vaccine should be given in children at 12 and 18 months of age. It should usually combined with MMR (measles, mumps and rubella).

Herpes Zoster

It is also called as '*shingles*' or '*zona*'. It is an acute infectious viral disease of extremely painful and incapacitating nature, characterized by inflammation of dorsal root ganglion, associated with vesicular eruptions of skin and mucous membrane of the area supplied by the affected sensory nerve.

Predisposing Factors

- **Trauma**—in some cases, trauma in the distribution of trigeminal nerve can lead to herpes zoster infection.
- **Malignancy**—development of malignancy or tumor in the region of dorsal root ganglion can also cause herpes zoster.
- **Radiation**—Local X-ray radiation can also be predisposing factors.
- **Immunosuppressive therapy**—this will lead to reactivation of virus and development of lesion.

Clinical Features

- **Age and sex distribution**—it affects adults and there is no sex predilection.
- **Prodromal symptoms**—prodromal period of 2 to 4 days in which shooting pain, paresthesia, burning and tenderness appears along the course of affected nerve. The reason for shooting pain is occurrence of active ganglionitis with resultant neuronal necrosis and severe neuralgia. The pain is present in the area supplied by the affected nerve (dermatome).
- **Appearance**—unilateral vesicles on an erythematous base appear in clusters, chiefly along the course of nerve and giving picture of a single dermatome involvement (**Fig. 31-12**). This is called as 'zosteriform' pattern.



Fig. 31-12: Intact vesicle seen showing dermatome involvement (Courtesy Dr Pincha).

- **Healing**—vesicle turns into scab in 1 week and healing takes place in 2 to 3 weeks.
- **Nerves**—nerves commonly affected are C₃, T₅, L₁, L₂ and 1st division of trigeminal nerve. It may affect motor nerve.
- **Involvement of 1st division of trigeminal nerve**—involvement of 1st division leads to corneal scarring and blindness (**Fig. 31-14**) which are presumably related to viral spread, neural damage, vasculitis and inflammatory immune response. 1st division involvement also lead to lesion of scalp (**Fig. 31-13**).



Fig. 31-13: Herpes zoster involving scalp of the patient.



Fig. 31-14: Involvement of ophthalmic division showing involvement of eyes (Courtesy Dr Revent Chole).

- **Zoster sine eruptione or zoster sine herpette**—it is zoster infection without appearance of dermatomal lesion.
- **Hutchinson sign**—it is cutaneous zoster infection of the side of tip of nose.

Oral Manifestations

- **Cause of oral lesion**—herpes zoster involving 2nd and 3rd divisions of trigeminal nerve lead to oral manifestation. Involvement of 2nd division leads to lesion of midface and upper lip and involvement of 3rd division leads to lesion of lower face and lower lip.
- **Site**—it may be found on buccal mucosa, tongue, uvula, pharynx and larynx.
- **Symptoms**—patient noticed pain, burning, tenderness usually on the palate on one side.
- **Signs**—after several days of symptoms, intact vesicles (**Fig. 31-15**) appear which soon rupture (**Fig. 31-16**) to leave areas of erosion (**Fig. 31-17**).



Fig. 31-15: Intact vesicle seen on the face by herpes zoster (Courtesy Dr Jitu Sachdeo).



Fig. 31-16: Herpes zoster showing ruptured vesicle in the oral cavity.



Fig. 31-17: Erosion area seen on the palate in case of herpes zoster (Courtesy Dr Chole).

- *Healing*—healing usually takes place within 10 to 14 days (**Fig. 31-18**).



Fig. 31-18: Healed herpes zoster lesion seen on the face (Courtesy Dr Jitu Sachdeo).

- *Teeth*—trigeminal herpes zoster occurring during tooth formation causes pulpal necrosis and internal root resorption. So in the case of herpes zoster, several non-vital teeth can be seen (**Fig. 31-19**). In some cases, osteomyelitis can also develop in the patient.

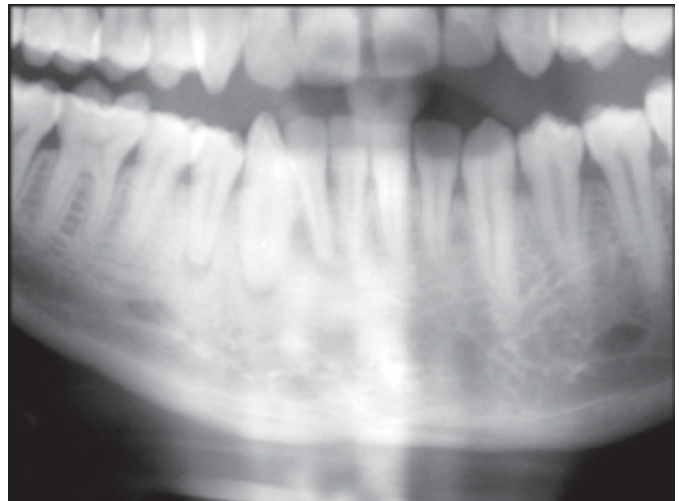


Fig. 31-19: Multiple non-vital teeth seen 7 years after occurrence herpes zoster (Courtesy Dr Chole).

Diagnosis

- *Clinical diagnosis*—lesions along the distribution of nerve will give clue to diagnosis.
- *Fluorescent antibody testing*—in this test, smear is obtained by scraping the lesion and staining it with fluorescent conjugated monoclonal antibody.

Table 31-1: Difference between herpes zoster and recurrent herpes simplex infection

<i>Herpes zoster</i>	<i>Recurrent herpes simplex infection</i>
Prodromal symptoms—fatigue, hyperesthesia, pain	Prodromal symptoms—tension, burning, itching
Development—edema and erythema, papulovesicular then vesiculopustular lesions and erosion	Development—vesiculoerosive lesion in crops and clusters, but not limited to dermatome
Severe pain	Moderate pain
Lesions limited to the course of a sensory nerve	No skin lesions
Longer course—post-zoster neuralgia	Fast healing without consequence
No recurrence	Recurrent appearance

- **PCR**—Polymerase chain reaction testing is used to detect viral antigen.
- **Biopsy**—it will show multinucleated giant cells.
- **Virus isolation**—this virus is difficult to culture.

Differential Diagnosis

- **Recurrent herpes simplex infection**—described in **Table 31-1**.
- **Herpangina**—acute infection, palatal vault is affected.

Management

- **Antiviral drugs**—acyclovir 800 mg five times daily which is associated with significantly accelerated healing within 48 hours of the onset of rash.
- **Symptomatic treatment**—antipyretic medication with antipruritics diphenhydramine can be administered to decrease itching.
- **Prevention of postherpetic neuralgia**—intralesional steroids and local anesthetic can be used to decrease healing time and to prevent postherpetic neuralgia. But this comes with many side effects and there are some conflicting reports about the efficiency of steroid in control of postherpetic neuralgia.
- **Capsaicin**—topical capsaicin 0.025% four times a day has been suggested for temporary relief of neuralgia following herpes zoster infection. Capsaicin is derived from red peppers. The mechanism of action apparently involves the depletion of substance P in the peripheral sensory neurons causing the skin less sensitive. After treatment patient should wash hand after use and avoid contact with mucosal surface.
- **Tetracycline rinse**—mouth rinsing with tetracycline, three to five times daily, may reduce the pain.

Postherpetic Neuralgia

This is complication of zoster infection.

Etiopathogenesis

- **Peripheral injury**—zoster virus injures the peripheral nerve by demyelination, wallerian degeneration and sclerosis.
- **Central injury**—there is also atrophy of dorsal horn cells in the spinal cord.
- **Infection**—low grade persistent infection of trigeminal ganglion also cause postherpetic neuralgia.

Clinical Features

- **Age and sex distribution**—It affects older age group. Incidence of postherpetic neuralgia is more in women as compared to men.
- **Symptoms**—initially, there is presence of rash which is followed by pain. Pain may continue for weeks to months. If pain occurs more than 6 months of duration is usually called as postherpetic neuralgia.
- **Other symptoms**—there may be paresthesia, hyperesthesia and allodynia. In some cases, there is also presence of sensory deficit.

Management

- **Prevention**—use of live attenuated Varicella zoster vaccine after age of 60 years will reduce the incidence of postherpetic neuralgia. To control postherpetic neuralgia, antiviral drug should be given in early course of disease.
- **Topical therapy**—use of topical agents like lidocaine (anesthetics), capsaicin (an extract of hot chili peppers that depletes the neurotransmitter substance P) should be used.
- **Drug therapy**—the use of TCA (tricyclic antidepressant) like Amitriptyline, nortriptyline, doxepin and desipramine have been used to minimize painful sequelae of this infection. The patient who cannot tolerate TCA due to cardiovascular side effect, the other drug gabapentin and pregabalin should be given. Carbamazepine or phenytoin can also be given to minimize the pain.

- **Surgery**—when drug therapy is ineffective then surgery should be carried out at the level of peripheral nerve or dorsal root is indicated. In some cases, sympathetic nerve block is also effective.
- **Steroid therapy**—steroid injection can be given in a patient with age more than 60 years, for the treatment of post-herpetic neuralgia. This will help in reducing the pain of the patient.

James Ramsay Hunt Syndrome

It is zoster infection of geniculate ganglion with involvement of the external ear and oral mucosa.

The clinical manifestation of it is facial paralysis as well as pain of the external auditory meatus and pinna of the ear.

In addition, vesicular eruption occurs in the oral cavity and oropharynx with hoarseness of voice, tinnitus, vertigo and occasional other disturbances.

Mumps

It is described in Chapter 26: Salivary Gland Disorders.

Measles

It is also called as '*Rubeola*' or '*morbilli*'. It is an acute contagious dermatotropic viral infection, primarily affecting children and occurs many times in epidemic form.

Transmission

- **Droplet infection**—spread of disease occurs by direct contact with a person or by droplet infection, the portal of entry being the respiratory tract.

Clinical Features

- **Incubation period**—incubation period is 8 to 10 days.
- **Prodromal symptoms**—onset of fever, malaise, cough and coryza (running nose).
- **Eye lesion**—there is conjunctivitis, photophobia, lacrimation occurs.
- **Skin lesion**—skin eruption begins on face, in the hair line and behind the ear and spread to neck, chest, back and extremities. They appears as tiny red macules or papules which enlarge and coalesce to form blotchy discolored irregular lesions, which blanch on pressure. Skin lesions fade away in 4 to 5 days with fine desquamation.
- **Complication**—it is the disease which lowers general body resistance and for this, it often leads to complications. Complications include bronchial asthma, encephalitis, otitis media, noma, and Hodgkin's lymphoma.

Oral Manifestations

- **Onset**—oral lesions precede 2 to 3 days before cutaneous rash and are pathognomonic of this disease.
- **Site**—the most common site is on buccal mucosa.
- **Koplik's spots**—intraoral lesions are called as Koplik's spots and occur in 97% of cases (**Fig. 31-20**). They are small, irregularly shaped flecks which appear as bluish white specks surrounded by bright red margins. They are described as '*grains of salt*' on red background.
- **Other features**—there is generalized inflammation, congestion, swelling and focal ulceration of gingiva, palate, and throat may occur.



Fig. 31-20: Bluish white numerous Koplik spots seen in measles.

Diagnosis

- **Clinical diagnosis**—typical clinical features like Koplik spots and history are important diagnostic criteria for measles.
- **Laboratory diagnosis**—it is done by virus isolation and raised antibody titers. Antibodies appear after 3 days of rash.

Differential Diagnosis

- **Small pox**—high fever, monoform exanthema.
- **Chickenpox**—typical exanthema that follows the intraoral lesion. The diagnosis can be established microscopically (blister swab) and virologically.

Management

- **Vaccination**—best treatment for the measles is vaccination. The widely used vaccine is MMR. One injection of live, attenuated measles virus should be given subcutaneously in children over one year. Second dose of vaccine is given in 12 to 15 months.
- **Passive immunization**—human immunoglobulin given intramuscularly is recommended for the prevention and attenuation of measles, particularly for contact under

18 months of age and for debilitated children. The dose is 250 mg for children under 1 year and 500 mg for those over this age.

- *Isolation of patient*—the patient should be isolated, if possible.
- *Symptomatic treatment*—analgesic, anti-pyretic should be given to patient to control pain and fever.
- *Specific drug treatment*—no drug has shown any promising results in the management of measles. Drugs which are tried are ribavirin, interferon, immunoglobulin and vitamin A.

Rubella (German Measles)

Rubella is produced by Togavirus. It is milder infection as compared to measles.

Transmission

- *Droplet infection*—it is transmitted through droplet infection in individual who is living in close living condition.
- *In utero*—infants with congenital infection may be present. It may result in birth defect. Infant born with rubella is called as *congenital rubella syndrome* (CRS).

Clinical Features

- *Incubation period*—incubation time is 14 to 21 days.
- *Prodromal symptoms*—it includes fever, headache, malaise, anorexia, conjunctivitis, coryza, cough and pharyngitis.
- *Lymphadenopathy*—it is seen in suboccipital, postauricular, and cervical chains.
- *Complication*—it includes arthritis, encephalitis and thrombocytopenia.
- *Rash*—the exanthematous rash seen on face and neck. It is discrete pink macule and it fades as it spreads.
- *Congenital rubella syndrome*—it results in deafness, heart disease and cataracts.

Oral Manifestation

- *Forschheimer's sign*—small, discrete, dark red papules seen on soft palate.
- *Petechiae*—petechiae can also be seen on the palate.

Diagnosis

- *Clinical diagnosis*—it is very difficult to establish clinical diagnosis as clinical presentation is mild and subclinical.
- *Laboratory*—serological analysis should be carried out to establish diagnosis.

Management

- *Vaccination*—this has dramatic results after vaccine was released in 1969.
- *Human rubella immunoglobulin*—it is administered to have passive immunity. If it is given within days after the exposure, it will decrease the severity of infection.

Coxsackievirus Infection

They are RNA retroviruses and are named after town in upper New York where they were first discovered. They are divided into 2 groups:

- Type A—24 types
- Type B—6 types

These viruses can cause hepatitis, meningitis, myocarditis, pericarditis and respiratory disease. Three clinical types of infection are important and discuss below. These diseases occur more frequently from June to October (summer and early fall).

Transmission

- *Fecal oral route*—it is major path of transmission. Frequent hand washing will diminish the spread of this virus.
- *Saliva*—virus can spread through saliva during acute condition.
- *Predisposing factors*—overcrowding, poor oral hygiene may aid to infection.

Herpangina

It is also called as '*aphthous pharyngitis*', '*vesicular pharyngitis*'. A₄ causes majority of the cases. A₄ to A₁₀ and A₁₆ to A₂₂ have also been implicated.

Clinical Features

- *Age*—majority affected are young children aged 3 to 10 years.
- *Incubation period*—incubation period is of 2 to 10 days.
- *Site*—it occurs on posterior pharynx, tonsil, faucial pillars and soft palate.
- *Prodromal symptoms*—initially, generalized symptoms of fever, chills, headache, anorexia, prostration, abdominal pain and sometimes vomiting. Sore throat, dysphagia and occasionally, sore mouth can occur.
- *Ulceration*—lesion starts as punctate macule which evolves into papules and vesicles (**Fig. 31-21**). Within 24 to 48 hours, vesicles get ruptured forming small 1 to 2 mm ulcers.
- *Base and margins*—ulcers show a gray base and inflamed periphery.
- *Healing*—they generally heal without treatment in 1 week.



Fig. 31-21: Ulceration seen on posterior part of oral cavity in case of herpangina.

Diagnosis

- *Clinical diagnosis*—lesions are seen posterior part of oral mucosa.

Differential Diagnosis

- *Primary herpes simplex infection*—herpangina occur in epidemic, and HSV does not. Clinical manifestations of herpangina are generally milder than HSV infection. Lesions of herpangina occur in pharynx and posterior portions of oral mucosa and in case of herpes simplex infection, it occur in anterior part of oral cavity. Herpangina does not cause generalized acute, marginal gingivitis which is typical feature of herpes simplex infection.
- *Herpes zoster*—in this, segmental distribution of vesicle is seen which is absent in case of herpangina.

Hand-Foot-and-Mouth Disease

It is caused by A₁₆, A₅, A₇, A₉, A₁₀, B₂ and B₅.

Clinical Features

- *Age*—it primarily affects children between the age of 6 months and 5 years.
- *Symptoms*—there is anorexia, low grade fever, diarrhea, nausea and vomiting.
- *Appearance*—it is characterized by appearance of maculopapular, exanthematous and vesicular lesions of skin, particularly involving the hands, feet, legs, arms and occasionally buttocks.

Oral Manifestations

- *Sites*—the most common sites for oral lesions are hard palate, tongue and buccal mucosa.
- *Symptoms*—a sore mouth with refusal to eat is one of the most common findings in this disease.

- *Appearance*—oral lesions are more extensive than herpangina. The tongue may become red and edematous. Clinical manifestations last for 3 to 7 days.

Diagnosis

- *Clinical diagnosis*—oral lesion in association with skin lesion will aid to diagnosis.

Differential Diagnosis

- *Herpetic gingivostomatitis*—entire oral cavity is affected. No hand and feet involvement.
- *Varicella zoster infection*—segmental distribution along the anatomical location of nerve (unilateral lesions).
- *Herpangina*—affect children mostly in late summer and early monsoon and lesions are common on soft palate and facial area with fever and malaise.
- *Allergic stomatitis*—sudden appearance, no prodromal symptoms with itching and noticeable erythema.
- *Chickenpox*—in addition to intraoral changes, polymorphous exanthema on the entire body and severe lesions.

Acute Lymphonodular Pharyngitis

It is caused by A₁₀ and is same as herpangina. Yellow-white nodules appear that do not progress to vesicles or ulcers. It is self limiting and only supportive care is indicated.

Clinical Features

- *Age*—the disease affects predominantly children and young adults, occasionally older adults can also be affected.
- *Site*—the lesion appears on uvula, soft palate, anterior pillars and posterior oropharynx.
- *Incubation period*—it has got 5 days incubation period and course may run for 4 to 14 days, with local oral lesions resolving within 6 to 10 days.
- *Symptoms*—the chief complain is of sore throat, 41°C temperature, mild headache, anorexia and loss of appetite.
- *Appearance*—it consists of raised, discrete, and whitish to yellowish solid papules of 3 to 6 mm in diameter, surrounded by narrow well defined zone of erythema.
- *Signs*—lesion is non-vascular, non-ulcerated, tender, superficial and bilateral.

Diagnosis

- *Clinical diagnosis*—whitish nodule with sore throat can lead to diagnosis.

Management

- *Symptomatic treatment*—no specific treatment is necessary since the disease is self limiting and generally

regresses within one to two weeks. Symptomatic treatment directed towards giving anti-pyretic and topical anesthetics.

- *Nutritional supplement*—as eating and swallowing is difficult, patient should be given proper hydration.

Infectious Mononucleosis

It is also called as 'glandular fever' or 'mono'. It is a benign acute infectious disease caused due to the Epstein-Barr virus, a herpes virus which infects the B-lymphocytes. It is commonly seen in adolescents and young adults. The mechanism of human transmission is not entirely known but one important mean is thought to be through deep kissing so this condition is also called as 'kissing disease'. EBV is present in oropharyngeal secretions and mixed saliva during active phase.

Clinical Features

- *Incubation period*—it varies from 10 to 40 days.
- *Age*—disease occurs chiefly in children and young adult in 4 to 15 age groups.
- *Symptoms*—the patient usually complaint of sore throat accompanied by fever usually 101°F to 103°F and extreme fatigability. Occasionally, there is a complaint of headache, photophobia, nausea, vomiting, diarrhea and presence of erythematous macular rash (*Morbili-form skin rash*).
- *Signs*—physical examination reveals enlarged palatine tonsils with copious amount of cheesy yellow exudate filling tonsillar crypt. Patient may also be having splenomegaly and hepatomegaly.
- *Lymphadenopathy*—enlargement of the superficial lymph nodes, particularly the posterior cervical are common manifestations. Lymph nodes are slightly tender on palpation.
- *Chronic fatigue syndrome*—this is controversial whether this is associated with EBV infection. In this, patient noticed chronic fatigue, fever, pharyngitis, myalgia, headache, arthralgia, paresthesia, depression and cognitive defect.
- *Complications*—airway obstruction, splenic rupture, progressive neurological involvement and hemolytic anemia.
- *Hematological finding*—there is an increase in white blood cell count.

Oral Manifestations

- *Site*—lesion present on soft palate, labial and buccal mucosa.
- *Petechiae*—it can be seen on soft palate (**Fig. 31-22**). They are transient.



Fig. 31-22: Multiple petechiae seen on soft palate in the patient of infectious mononucleosis.

- *Ulcerative gingivitis, periodontitis and stomatitis*—ulcerative gingivitis, periodontitis and stomatitis may be present and the lesion normality persisting for 3 to 11 days.
- *Tonsils*—intraorally, the most prominent sign is enlargement and inflammation of the tonsils along with sore throat and difficulty in swallowing. Quite commonly the tonsils are covered by a white or grayish pseudo-membrane.
- *Bleeding*—1/3rd of the patients with hemorrhagic tendency exhibit oronasopharyngeal bleeding, including bleeding from gingiva.

Diagnosis

- *Clinical diagnosis*—morbiliiform skin rash with enlarged palatine tonsil with fever can aid in diagnosis.
- *Laboratory diagnosis*—there is positive Paul Bunnell (test for detecting EBV antibody) and mono spot test.

Management

- *Symptomatic*—the oral lesion of infectious mononucleosis is treated symptomatically. A topical anesthetic may be used in painful ulcers and hydrogen peroxide rinses aid in ameliorating the fusospirochetal gingivitis. Patient should be given antipyretic and anti-inflammatory therapy to control fever and pain.
- *Antiviral drugs*—ganciclovir and Alfa interferon inhibit the replication of Epstein Barr virus *in vitro*. Alpha interferon reduces EBV replication and shedding in organ transplant patient.
- *Corticosteroid*—corticosteroids are indicated only in the presence of complications like airway obstruction, progressive neurological involvement, hemolytic anemia. Normally corticosteroid should be avoided.
- *Precaution*—patient should avoid sports as it may lead to splenic rupture.

Cytomegalovirus Infection

This virus is also called as HHV-5. It may be clinically expressed or latent. CMV can remain latent in salivary gland cells, endothelium, macrophages and lymphocytes. Cytomegalovirus (CMV) is a double-stranded DNA virus that is fairly common in the general population, with approximately 60% of people being seropositive but asymptomatic.

Transmission

- *In utero transmission*—infants can acquire it *in utero* from placenta, during delivery. Infant can also acquire infection through breastfeeding.
- *Sexual transmission*—transmission can also occur through exchange of body fluid like saliva, during sexual activity.
- *Blood transfusion and organ transplantation*—blood and transplanted tissues are also potential means of transmission of virus to susceptible individuals.

Clinical Features

- *Neonatal infection*—features of this infection include hepatosplenomegaly, extramedullary cutaneous erythropoiesis, encephalitis and thrombocytopenia. Encephalitis may lead to mental retardation and motor retardation.
- *Adult's infection*—there is fever, malaise, myalgia, abnormal liver function. Petechial hemorrhages can also occur. Patient may also suffer from pneumonia, microcephaly, cerebral calcification and hearing defect.
- *CMV chorioretinitis*—this is commonly seen in AIDS patient. This may lead to blindness of patient.
- *CMV colitis*—this will lead to bloody diarrhea.

Oral Manifestations

- *Salivary gland involvement*—this can occur in immunocompetent patient. Patient usually present with acute sialadenitis which can involve major and minor salivary gland. Affected gland is painful. Patient may suffer from xerostomia.
- *Mucosal ulceration*—oral ulcer occur in patient with CMV disease. It is more commonly seen in AIDS patient.
- *Gingival infection and hyperplasia*—patient may suffer from gingivitis and gingival hyperplasia (**Fig. 31-23**).
- *Dental significance*—susceptible dental personnel may acquire CMV infection in the absence of barrier protection.

Diagnosis

- *Clinical diagnosis*—CMV produces deep, penetrating oral ulcerations on the lips, tongue, pharynx, or any mucosal site.



Fig. 31-23: Gingival infection and ulceration seen in the patient with cytomegalovirus infection.

- *Laboratory diagnosis*—characteristic “owl’s eye” appearance of cellular inclusions during the histologic examination.

Management

- *Prevention*—it should be done by passive immunization with hyperimmune gamma globulin.
- *Antiviral therapy*—no specific antiviral therapy is useful. Foscarnet, ganciclovir and alpha interferon can be used which may yield success in some cases. Intravenous ganciclovir will produce resolution of oral ulceration in most instances.

Foot and Mouth Disease

It is also called as ‘aphthous fever’, ‘hoof and mouth disease’, ‘epizootic stomatitis’. It is a viral infection which rarely affects man, but does affect hogs, sheep as well as cattle. Transmission of this disease occurs through infected animals in human beings, it is usually through milk from infected animals.

It is manifested by fever, nausea, vomiting, malaise and appearance of ulcerative lesions of oral mucosa and pharynx. There is development of vesicle on skin. In some cases, they occur usually on the palms of hands and soles of feet.

Intraorally it can occur at any site, but lips, tongue, palate and oropharynx appear to be affected. These lesions being as small vesicles which rapidly rupture, but heal within two weeks.

Condyloma Acuminatum

It is also called as ‘genital wart’, ‘venereal wart’ or ‘verruca acuminata’. It is caused by human papillomavirus (HPV). It can be transmitted from mother to infant, at birth and resulting syndrome is called as *juvenile onset respiratory papillomatosis*.

Clinical Features

- *Incubation period*—it is about 1 to 3 months from the time of sexual contact.
- *Locations*—it is seen on external genitals, perianal mucosae, and adjacent skin as well as in vaginal and anal canal. Wart growth is favored by moist warm environment of perianal skin and mucosal surface.
- *Appearance*—they are a pink, fleshy papillomatous lesion which proliferate and coalesce rapidly to form diffuse papillomatous clusters of varying sizes.

Oral Manifestations

- *Sites*—it may involve gingiva, cheek, lip, hard palate, tongue and floor of mouth.
- *Appearance*—small keratotic warts occurring alone or in clusters on oral mucosa can be seen.
- *Size*—the enlargement may be larger than 1 cm in diameter.
- *Margins and base*—the lesion is sharply delineated and may appear sessile and pedunculated.

Diagnosis

- *Clinical diagnosis*—sessile pink pedunculated growth with history of oral sex is important in clinical diagnosis.
- *Laboratory diagnosis*—virus isolation can be done by staining of viral antigen DNA by hybridization restriction, endonuclease analysis and polymerase chain reaction.

Differential Diagnosis

- *Focal epithelial hyperplasia*—in it, fine granular surface texture and plaque-like shape of enlargement while in case of condyloma, there is cauliflower appearance.

Management

- *Surgical*—genital warts are treated by excision, electro- or cryosurgery, CO₂ laser therapy.
- *Application of chemical agents*—application of chemical agents such as podophyllin, cantharidin and 5-fluorouracil can be given.
- *Immunomodulating agents*—immunomodulating agent such as interferon can also be used in case of condyloma acuminatum.

Verruca Vulgaris (Common Wart)

Verruca vulgaris is caused by HPV virus type 2,4,6 and 40. It is transmitted by autoinoculation and develops on other part of the body and skin.

Clinical Features

- *Age*—it is common in children and some cases may be found in middle age.
- *Site*—skin of hand is common site and in case of oral cavity it is commonly present a vermilion border, labial mucosa and anterior tongue.
- *Appearance*—it appear as papular or nodular growth with papillary projection. The color of cutaneous lesion is pink, yellow, or white and oral lesions are usually white in color (**Fig. 31-24**).
- *Surface of lesion*—rough and pebbly.
- *Base*—base is pedunculated or sessile.
- *Keratin horn or cutaneous horn*—in some cases, extreme accumulation of keratin will result in hard surface projection. This is called as cutaneous horn or keratin horn.



Fig. 31-24: Common wart seen on finger (Courtesy Dr Pincha).

Diagnosis

- *Clinical diagnosis*—keratin horn with pedunculated surface pebbly lesion on hand will diagnose this condition.
- *Laboratory diagnosis*—there is proliferation of hyperkeratotic stratified squamous epithelium which are arranged in finger-like projection. There is also cupping effect.

Management

- *Cryotherapy*—liquid nitrogen cryotherapy is the treatment of choice in this condition. Cryotherapy will induce subepithelial blister which lifts the infected epithelium from the connective tissue allowing it to slough away.

- *Surgical excision*—surgical excision is usually carried out in case of oral lesion.
- *Keratinolytic agents*—topical application of keratinolytic agents can also be done.

Molluscum Contagiosum Infection

It is caused by molluscum contagiosum virus pox group.

Transmission

- *Skin contact*—it occurs due to intimate skin contact.
- *Sexual contact*—there is marked increase at the time of onset of sexual activity.
- *Predisposing factors*—poverty, overcrowding and poor hygiene are predisposing factors for transmission of virus.

Clinical Features

- *Incubation period*—incubation period is 14 to 50 days.
- *Age*—it is more common in children and young adults.
- *Sites*—it is more common on skin of inner thigh, lower abdomen or external genitals. Some cases may be seen on lips, tongue and buccal mucosa.
- *Appearance*—it manifests as multiple or isolated discrete elevated nodules, or sometimes papules, with depressed centers, which may be keratinized and are normal or pink in color.
- *Adjacent area*—adjacent area is surrounded by the zone of erythema.
- *Shape and size*—these lesions are hemisphere in shape, usually about 5 mm in diameter.
- *Number*—multiple lesions sometimes numbering 100 can also occur.
- *Healing*—it is self limiting and regresses spontaneously within 1 to 2 months.

Diagnosis

- *Clinical diagnosis*—discrete nodule with depressed center is typical of this disease.
- *Laboratory diagnosis*—it shows large eosinophilic intracytoplasmic inclusion bodies known as Henderson-Paterson inclusion or simply Molluscum bodies, measuring approximately 25 microns in diameter in the biopsy.

Management

- *Surgical therapy*—it includes curettage, followed by local cautery, cryotherapy.
- *Topical therapy*—topical application of caustic acid and irritants such as phenol, TCA, podophyllin and cantharidin.

Epstein-Barr Virus (EBV) Infection

The prevalence of EBV antibodies increase in adolescence and early adulthood due to increased opportunity for exchange of EBV in saliva, associated with adolescent, social activity. In this group, it is often characterized by a syndrome referred as infectious mononucleosis or '*kissing's disease*'. EBV may occur as blood borne infection.

EBV is associated with:

- Infectious mononucleosis (discussed in Chapter 37: Blood Disorders)
- Anaplastic nasopharyngeal carcinoma
- Burkitt's lymphoma (discussed in Chapter 37: Blood Disorders)
- B cell lymphoma (discussed in Chapter 37: Blood Disorders)
- Hairy leukoplakia (discussed in Chapter 35: AIDS).

Smallpox

It is also called as '*variola*'. On December 9, 1979, the WHO global commission for the certification of smallpox declared that smallpox eradication has been achieved throughout the world. Thus, there has been for the first time in history that a disease was totally eliminated from this planet.

Previously this disease manifested as occurrence of high fever, nausea, vomiting, chills and headache. The patient is extremely ill and may become comatose during this period. Orally there is ulceration of oral mucosa and pharynx is common. Multiple vesicles appear and rupture to form ulcers of non-specific nature.

Immunization is essential in the early period of life.

Chikungunya

Chikungunya is a relatively rare form of viral fever caused by an alpha virus that is spread by mosquito bites from the *Aedes Aegypti* mosquito, though recent search by the Pasteur Institute in Paris claims the virus has suffered a mutation that enables it to be transmitted by *Aedes Albopictus* (Tiger mosquito). Chikungunya is closely related to O'nyong'nyong virus.

The disease was first described by Marion Robinson and W.H.R. Lumsden in 1955, following an outbreak on the Makonde Plateau, along the border between Tanganyika and Mozambique, in 1952.

Chikungunya is not considered to be fatal. However, in 2005-2006, 200 deaths have been associated with this disease on Reunion Island and a widespread outbreak in southern India. Tamilnadu reported the largest no. of cases as of July 2006, specifically in Madurai and Tirunelveli.

Clinical Features

- **Symptoms**—it includes fever which can reach 39 degree Celsius. Fever lasts for 2 days and comes down. There is petechial or maculopapular rash usually involving the limbs and trunk, and arthralgia or arthritis affecting multiple joints which can be debilitating.
- **Other symptoms**—there can also be headache, conjunctival infection and slight photophobia.
- **Persistent symptoms**—however, joints pain, intense headache, insomnia and extreme degree of prostration lasts for about 5-7 days.
- **Dermatological manifestation**—it includes maculopapular rash, nasal blotchy erythema, lymphoedema over acral area, multiple ecchymotic spots, vesiculobullous lesions, sublingual hemorrhage, photo urticaria and acral urticaria
- **Oral manifestation**—lichenoid eruptions, aphthous ulcers (**Fig. 31-25**), freckle pigmentation over facial area.



Fig. 31-25: Aphthous type ulceration seen in Chikungunya patient.

Management

- There is not any treatment available for this disease. Only symptomatic treatment in the form of analgesics can be given.

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Introduction

Nowadays as patient is receiving broad spectrum antibiotics, radiation, cytotoxic and immunosuppressive drugs, there is increases incidence of fungal disease in the society. Many of the fungal disease are not diagnosed due to lack of diagnostic facility. Fungi belongs to plant kingdom which are non-photosynthetic due to absence of chlorophyll. Pathogenic fungi exists in three forms like yeasts, yeast like and filamentous organism.

Infection Caused by Fungi

The spectrum of infection cause by fungi is wide and there is no satisfactory classification of fungal disease. Some important dental point of view fungal disease are described below.

Histoplasmosis

It is also called as 'Darling's disease'. It is caused by *Histoplasma capsulatum*, a dimorphic fungus that grows in the yeast form in infected tissue. Infection results from inhalation of dust contaminated with dropping, particularly from infected birds.

Types

- Acute primary histoplasmosis
- Progressive disseminated histoplasmosis
- Chronic cavitory histoplasmosis.

Clinical Features

- *Acute primary histoplasmosis*—primary infection is mild, manifesting as self limited pulmonary disease that heals to leave fibrosis and calcification. There is chronic low grade fever, malaise, headache and productive cough. There may be pleuritic pain. Chest radiograph may

show patchy infiltrate which may exhibit signs of calcification.

- *Progressive disseminated histoplasmosis*—it is seen in children and elders. It is manifested in hepatosplenomegaly and lymphadenopathy. Patients with disseminated form show evidence of bone marrow involvement by anemia and leukopenia. There is also kidney involvement with gastrointestinal and oropharyngeal ulcerative lesions.
- *Chronic cavitory histoplasmosis*—it closely mimics chronic cavitory tuberculosis. The cavitory lesions are bilateral and are found in the upper lung fields. Symptoms include cough, dyspnea and weight loss.
- *Laboratory diagnosis*—the organisms are found in large numbers in phagocytic cells and appear as tiny intracellular structures measuring little more than 1 micron in diameter. Biopsy shows small oval yeasts within macrophages and reticuloendothelial cells as well as chronic granuloma, epithelial cells, giant cell with caseation necrosis.

Oral Manifestations

- *Sites*—it is seen on buccal mucosa, gingiva, tongue, palate or lip. Oral lesions are common in the progressive disseminated form.
- *Symptoms*—patient may complain of sore throat, painful chewing, hoarseness, difficulty in swallowing.
- *Appearance*—oral lesions are nodular, ulcerative or vegetative. If left untreated, it will progress to form firm papule or nodules which ulcerate and slowly enlarge. It resembles malignancy.
- *Base and surface*—ulcerated area covered by nonspecific gray membrane (**Fig. 32-1**) and is indurated.

Diagnosis

- *Clinical diagnosis*—hepatomegaly, splenomegaly, with oral ulcerative lesion will give clue to the diagnosis.



Fig. 32-1: Ulcerative lesion seen in female patient in histoplasmosis. It is mistaken as malignancy of mandibular vestibule.

- **Laboratory diagnosis**—Histoplasma organisms are found in phagocytic cells.

Differential Diagnosis

- **Tuberculosis**—sputum examination, tuberculin test.
- **Blastomycosis**—biopsy and culturing the organism from tissue.
- **Mucormycosis**—biopsy.
- **Cryptococcosis**—organisms cultured on Sabouraud's glucose agar.

Management

- **Supportive treatment**—patient should be given analgesic and antipyretics to control pain and fever.
- **Ketoconazole**—it should be given for 6 months in non-immunosuppressed patient.
- **Itraconazole**—this agent is more effective and less toxic than ketoconazole.
- **Amphotericin B**—intravenous amphotericin B should be given in chronic and severe form.

African Histoplasmosis

It is caused by fungus *Histoplasma duboisii* that is larger than *Histoplasma capsulatum*. The disease, because of its clinically distinct form and geographical distribution is called as African histoplasmosis.

The disease exists in two forms, i.e. localized and disseminated.

Clinical Features

- **Localized form**—it is confined to the skin, lymph nodes or localized bony areas. Bones when involved, may show cyst-like lesions.

- **Disseminated form**—in disseminated form, multiple cutaneous lesions and involvement of viscera, especially liver and spleen occurs. The course of disease is progressive and fatal.

Oral Manifestation

- **Site**—it involves mucosa and jaw bones.
- **Signs**—mandibular and maxillary involvement is usually presented as bony resorption, resulting in loosening of teeth.

Diagnosis

- **Clinical diagnosis**—multiple cutaneous lesion with loosening of teeth may give clue to diagnosis.

Management

- **Amphotericin B**—amphotericin B is the drug of choice and lesion may require surgical intervention.

Blastomycosis

It is caused by '*blastomyces dermatitidis*'. Organism is a normal inhabitant of soil and that is the reason for it to be common in agricultural worker. It is transmitted through the respiratory tract. Infection with blastomycosis begins in a vast majority of cases by inhalation. Oral lesion may be primary (local inoculation) or secondary (extrapulmonary dissemination) to some infection elsewhere in the body.

Types

- Primary pulmonary blastomycosis
- Cutaneous blastomycosis
- Disseminated or systemic blastomycosis.

Clinical Features

Primary pulmonary blastomycosis

- **Age and sex**—it is more common in men than women and typically occurs in middle age.
- **Symptoms**—it follows a chronic course with malaise, high grade fever and mild cough. If untreated, shortness of breath, weight loss and blood tinged sputum are encountered. In some cases, it may precipitate adult respiratory distress syndrome.

Cutaneous Blastomycosis

- **Sites**—infection of skin, mucosa and bone may also occur, resulting from metastatic spread of organism through lymphatic system.
- **Appearance**—skin and mucosal lesion starts as subcutaneous nodule and progresses to well circumscribed indurated ulcer.

- **Signs**—elevated, verrucous, crusted and single or multiple lesions developing on exposed part of the body, particularly on hands and face.
- **Margins**—lesion leaves a slanted serpiginous border with a tendency towards healing in the center. When the crust is lifted, pus exudes.

Disseminated or Systemic Blastomycosis

- **Cause**—it results from the spread of infection from pulmonary form.
- **Sites**—bones are involved in larger number of cases. Liver, kidney, spleen and gastrointestinal tract may be involved.
- **Signs**—cerebral abscess are found, which occur as a result of brain involvement.

Oral Manifestations

- **Symptoms**—oropharyngeal pain is present.
- **Signs**—enlargement of cervical lymph nodes occurs.
- **Appearance**—non-specific, painless verrucous ulcer with indurated borders is present. It is often mistaken for squamous cell carcinoma (**Fig. 32-2**). Some lesions are hard nodules and appear as sessile, granulomatous appearing plaque.



Fig. 32-2: Blastomycosis presenting as ulcerative lesion resembling squamous cell carcinoma.

Radiographic Features

- **Periosteal reaction**—radiograph may show periostitis and sub-periosteal new bone formation. Osteoblastic reaction is usually present in the later stages of disease.
- **Chest radiograph**—chest radiograph show concomitant pulmonary involvement in most of the cases.

Diagnosis

- **Clinical diagnosis**—the index of suspicion is increased when chronic, painless, oral ulcer appears in an agri-

cultural worker or when review of system reveals pulmonary symptom.

- **Radiological features**—chest radiograph will show pulmonary involvement.
- **Laboratory diagnosis**—diagnosis is made on the basis of biopsy and on culturing the organism from tissue.

Differential Diagnosis

- **Squamous cell carcinoma**—present for weeks or months, palpation shows induration, most common in older patient.
- **Tuberculosis**—undermined flabby borders, usually painless, sputum examination, Mantoux test.
- **Histoplasmosis**—culture and biopsy should be done.
- **Mucormycosis**—biopsy.
- **Cryptococcosis**—organism culture should be done.

Management

- **Itraconazole**—it is generally recommended for the patient of chronic blastomycosis.
- **Amphotericin B**—it is usually given in chronic case of blastomycosis. Intravenous amphotericin B for 8 to 10 weeks causes resolution of the disease. In case of acute blastomycosis, it does not require any treatment.

Mucormycosis

It is also called as '*phycomycosis, zygomycosis*'. It is caused by saprobic organism of class Zygomycetes. It is more common in patients with decreased resistance, due to diseases like diabetes, tuberculosis, renal failure, leukemia, cirrhosis and in severe burn cases. It begins with inhalation of fungus by susceptible individual.

Types

- **Superficial**—it involves external ear, fingernails and skin.
- **Visceral**
 - **Pulmonary**
 - **Gastrointestinal**
 - **Rhinocerebral or rhinomaxillary form**—it is important with dental point of view.

Clinical Features

- **Locations**—infection usually arise in lateral wall of nose and maxillary sinus; may rapidly spread by arterial invasion to involve the orbit, palate, maxillary alveolus and ultimately the cavernous sinus and brain through hematogenous spread and may cause death.
- **Symptoms**—ptosis, proptosis, nasal obstruction, fever, swelling of cheek and paresthesia of face can occur.

- *Intracranial involvement*—intracranial involvement may be heralded by cranial neuropathy, especially of the trigeminal and facial nerve. There is increased lethargy, progressive neurologic deficit and ultimately death.
- *Artery involvement*—fungus invades artery to cause fibrosis and ischemia of the supplied area.
- *Nose*—there is appearance of a reddish black nasal turbinate and septum. Nasal discharge caused by necrosis of nasal turbinate.

Oral Manifestations

- *Site*—most common site involved is maxillary sinus. Other sites which can also be involved are palate, lip, alveolar bone and gingiva.
- *Signs*—ulceration of palate may occur due to necrosis and invasion of palatal vessels. It is large and deep, causing denudation of underlying bone (**Fig. 32-3**). Ulcer may be seen on gingiva, lip and alveolar bone. Ulcer on palate may appear black and necrotic.
- *Massive destruction*—it can be seen when the condition is not treated at the earliest.



Fig. 32-3: Mucormycosis showing deep ulceration of the palate (Courtesy Dr Ashok L).

Radiographic Features

- *Appearance*—paranasal sinus may reveal mucoperiosteal thickening of the involved sinus. With disease progression, there is increased nodularity and soft tissue thickening, usually mimics a tumor on radiographical examination.
- *CT scan features*—CT scan is most helpful for detecting the degree of bone destruction and for evaluating disease extension into the orbit and brain (**Fig. 32-4**).

Diagnosis

- *Clinical diagnosis*—the nose involvement with destruction of palate, neurological finding may give clue to the diagnosis.

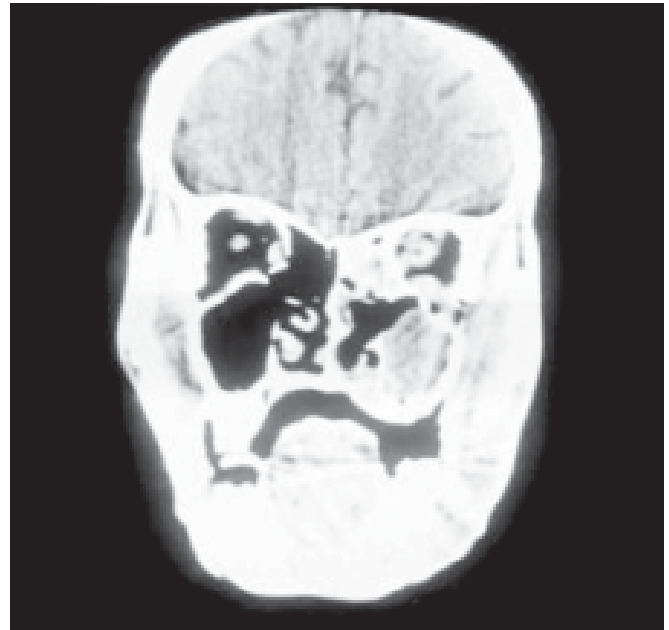


Fig. 32-4: CT scan showing increased radiopacity due to soft tissue mass in left maxillary sinus (Courtesy Dr Iswar).

- *Radiological finding*—CT scan will help in the diagnosis.
- *Laboratory diagnosis*—organism appear as nonseptate hyphae with branching at obtuse angles.

Differential Diagnosis

- *Squamous cell carcinoma*—indurated, longer history, resistance to therapy, firm borders, older patient, biopsy.
- *Necrotizing sialometaplasia*—rare, limited to hard and soft palate, usually painless.
- *Aphthous ulcer*—short duration, painful, heals in one to three weeks.

Management

- *Surgical debridement*—radical surgical debridement of infected and necrotic tissue should be carried out.
- *Amphotericin B*—systemic administration of high dose of amphotericin B should be given.
- *Control of underlying disease*—control of predisposing factors such as diabetes should be attempted.
- *Obturation*—prosthetic obturation of palatal defect should be carried out.
- *Symptomatic treatment*—it should be given to eliminate secondary infections and pain relief.

Cryptococcosis

It is also called as 'torulosis'. It is a chronic fungal infection caused by *Cryptococcus neoformans* and *Cryptococcus bacillispora*. The causative organism is gram positive, budding, yeast-like cell with an extremely thick, gelatinous capsule, measuring 5 to 20 microns in diameter. Infection

occurs due to inhalation of airborne microorganism. It has increased incidence in immunosuppressive patients, most commonly in HIV positive patient.

Clinical Features

- *Age*—there is slight predilection for middle aged males.
- *Site*—the infection usually occurs in lungs.
- *Symptoms*—it may be asymptomatic and in some cases, patient may complain of cough with mucoid expectoration. Occasionally, pleuritic pain and hemoptysis can also occur.
- *Skin lesion*—the skin lesion appears as multiple brown papules which ultimately ulcerate, the clinical picture is nonspecific.
- *Neurological features*—meningoencephalic lesions produce a variety of neurologic signs and symptoms, generally associated with increased intracranial pressure.
- *Coin lesion*—the radiograph of chest shows infiltrates and occasionally 'coin' lesion.
- *Diagnosis*—the organisms can be cultured on Sabouraud's glucose agar.

Oral Manifestations

- *Locations*—lesion of hard palate, soft palate, gingiva, extraction socket, tongue and tonsillar pillar are common.
- *Appearance*—they appear as simple non-specific, single or multiple ulcers. They are nodular and granulomatous, which may ulcerate during the course of disease.

Diagnosis

- *Clinical diagnosis*—the brown skin lesion with neurological features and non-specific ulcer seen in oral cavity.
- *Laboratory diagnosis*—gram positive yeast-like cell with gelatinous capsule.

Management

- *Ketoconazole*—mild to moderate cases can be treated with ketoconazole for 6 to 12 weeks.
- *Amphotericin B*—the severe form requires amphotericin-B, intravenously for up to 10 weeks.
- *Combination therapy*—combination therapy of amphotericin B and flucytosine is used in many cases to treat the disease. Another combination of fluconazole and itraconazole is also useful in Cryptococcosis infection.

Coccidioidomycosis

It is also called as 'valley fever', 'desert fever' or 'coccidioidal granuloma'. The disease appears to be transmitted to man and animals by inhalation of dust contaminated by the

spores of the causative organism, *Coccidioides immitis*. Symptoms occur usually 14 days after the inhalation of fungus. Infection is common in summer months, especially after periods of dust storm. It is self limiting and runs its course within 10 to 14 days. Lesions of head and neck, including the oral cavity, occur with some frequency.

Types

- Primary non-disseminated coccidioidomycosis.
- Progressive disseminated coccidioidomycosis.

Clinical Features

- *Age and sex*—it is common in all age groups and predominantly seen in males.
- *Primary pulmonary coccidioidomycosis*—the patient generally develops manifestations suggestive of respiratory disease such as cough, pleural pain, headache and anorexia. Patient may also complain of low grade fever and joint pain.
- *Primary cutaneous coccidioidomycosis*—skin lesions are also present, like erythema nodosum or erythema multiforme. Primary lesions, when they occur, are associated with regional lymphadenopathy. Granulomatous, verrucous or necrotic ulcers exuding thick pus are seen on involved skin surface.
- *Progressive disseminated coccidioidomycosis*—the disease usually runs rapid course and the dissemination extends from the lungs to various viscera, bones, joints, skin and central nervous system, where meningitis is the most frequent cause of death.

Oral Manifestations

- *Appearance*—the lesions of oral mucosa and skin are proliferative, granulomatous and ulcerated lesions that are non-specific in their clinical appearance.
- *Healing*—these lesions tend to heal by hyalinization and scar formation.
- *Lytic lesion*—lytic lesions of jaw may develop.

Diagnosis

- *Clinical diagnosis*—symptoms suggestive of respiratory disease, granulomatous and ulcerated oral lesion may give clue to the diagnosis.
- *Laboratory diagnosis*—biopsy shows mononuclear cell, lymphocytes, and plasma cells with foci of coagulation necrosis.

Management

- *Amphotericin B*—it has been found to be an effective chemotherapeutic agent for the disease. It is given in

patient who is immunosuppressive, disseminated infection and severe pulmonary infection. Long term therapy is required for complete cure.

- *Fluconazole*—it is given in high doses for extended period of time.
- *Ketoconazole*—it is used as an alternative therapy to amphotericin B.

Paracoccidioidomycosis

It is also called as 'South American blastomycosis', 'Lutz disease'. It is caused by *Paracoccidioides brasiliensis*. It is more commonly seen in farmers.

Clinical Features

- *Geographic location*—this is more commonly seen in South America.
- *Age and sex distribution*—it is exclusively found in males as compared to females. It is more common in middle age group.
- *Signs*—patient usually present with signs of pulmonary infection. In some cases, adrenal involvement also occur resulting in hypoadrenocorticism.

Oral Manifestation

- *Site*—commonly affected site are alveolar mucosa, gingiva, and palate. Lips can also be affected.
- *Appearance*—oral lesion appear as mulberry-like ulceration.

Diagnosis

- *Clinical diagnosis*—signs of pulmonary infection, with mulberry-like ulceration.
- *Laboratory diagnosis*—size of this organism is larger than North American form.

Management

- *Sulfonamide*—this can be used to treat mild to moderate infection.
- *Amphotericin B*—this is used in severe cases. It should be used intravenously.
- *Itraconazole and ketoconazole*—this can also be used in some cases when the condition is not life threatening.

Geotrichosis

It is caused by organism *Geotrichum candidum*. It is found in patients with debilitating diseases.

Clinical Features

- *Location*—these are more commonly found in lungs and oral mucosa.

- *Symptoms*—lung involvement produce symptoms of pneumonitis or bronchitis. The expectoration is often tinted with blood.

Oral Manifestations

- *Appearance*—they are similar to candidiasis or thrush (Fig. 32-5), being white, velvety, patch like covering of the oral mucosa, isolated or diffuse in distribution.
- *Tonsillar lesion*—tonsillar lesions are common in association with oral lesions.



Fig. 32-5: Geotrichosis presenting as candidiasis type lesion on oral cavity.

Diagnosis

- *Clinical diagnosis*—lung involvement with candidal type lesion in debilitated patient.
- *Laboratory diagnosis*—the organism is small, rectangular shaped; spores measuring approximately 4 to 8 microns, often with rounded ends. The tissue reaction is non-specific and of acute inflammatory type.

Management

- *Antifungal therapy*—it includes topical and systemic application of nystatin and amphotericin B.

Sporotrichosis

It is a fungal infection caused by *Sporotrichum schenckii*. The disease predominantly affects skin.

Causes

- *Exposure to animal*—exposure to a wide variety of animals, both domestic and wild.

- *Accidental injury*—accidental injury from the thorns of some plants or bushes. Accidental laboratory or clinical inoculation in hospital workers.

Clinical Features

- *Incubation period*—the incubation period is from a week to 3 weeks. It involves the skin, subcutaneous tissues and oral, nasal and pharyngeal mucosa.
- *Skin lesion*—the skin lesion often described as sporotrichotic 'chancre' appears at the site of inoculation as firm, red to purple nodule which soon ulcerates.
- *Lymph nodes*—regional lymphadenopathy is generally developed and it may ulcerate and drain.

Oral Manifestations

- *Location*—non-specific ulceration of the oral, nasal and pharyngeal mucosa.
- *Appearance*—long standing lesions become granulomatous, vegetative or papillomatous.
- *Symptoms*—pain is present and the cervical lymph nodes are always enlarged.

Radiographic Features

- *Incidence*—it rarely involves the bone, it resembles to those which occur in tuberculosis.
- *Appearance*—in the mandible, there is large destructive lesion in the molar region and ramus. It has a loculated cystic appearance, which causes marked expansion of the inferior aspect of the jaw.
- *Periosteal reaction*—in long bones, sporotrichosis may produce some periosteal new bone, but this reaction is confined to jaws of children.

Diagnosis

- *Clinical diagnosis*—lymph nodes enlargement with non specific ulceration of oral cavity will give clue to the diagnosis.
- *Radiological diagnosis*—periosteal reaction with expansion of inferior portion of jaw.
- *Laboratory diagnosis*—the fungus is small, ovoid branching organism with septate hyphae showing budding form. It is only 3 to 5 microns in diameter and can be cultured on Sabouraud's glucose agar medium. The tissue reaction is a granulomatous.

Management

- *Potassium iodide*—it includes oral administration of potassium iodide in suitable doses.

Rhinosporidiosis

It is caused by *Rhinosporidium seeberi*. It affects oropharynx and nasopharynx.

Clinical Features

- *Location*—it is more common on oropharynx, nasopharynx, larynx, skin, eyes and genital mucosa.
- *Symptoms*—initial symptoms include nasal irritation, accompanied by mucoid discharge.
- *Skin lesion*—the skin lesion appears as small verrucae or warts, which ultimately become pedunculated.
- *Signs*—posteriorly, these polypoid masses may extend into the pharynx. The lesions are soft, friable and highly vascular.
- *Genital lesions*—genital lesion resembles condyolomas.

Oral Manifestations

- *Sites*—the soft palate appears to be the most frequent site of oral involvement.
- *Appearance*—it is accompanied by a mucoid discharge and appears as soft, reddish pink, polypoid growth of tumor like nature which spreads to the pharynx and larynx. The lesions are vascular and bleed readily.

Diagnosis

- *Clinical diagnosis*—pedunculated wart with nasal polypoid mass, with intraoral soft reddish pink growth on soft palate.
- *Laboratory diagnosis*—the organisms appear as sporangia containing large number of round or ovoid endospores with size of 5 to 7 microns in diameter.

Management

- *Surgical*—surgical removal of the growths and application of cautery is the treatment recommended for rhinosporidiosis.

Aspergillosis

It is a fungal infection caused either by sensitization to parasitic colonization, or tissue invasion by species of genus *Aspergillus*. It is second to candidiasis, as an opportunistic infection, in immunocompromised patients. It is more commonly seen in diabetes mellitus patients.

Clinical Features

- *Sites*—the respiratory tract, external auditory canal, nasopharynx, cornea, gastrointestinal tract and occasionally the skin may be the primary sites of infection.

- *Symptoms*—in some cases, there may be asthmatic episode in sensitive persons.
- *Nasal aspergillosis*—it may present swelling, ulceration, crusting and necrosis of anterior turbinates, nasal septum and nasal wall.
- *Allergic fungal sinusitis*—in some cases, disease may appear as allergic infection appearing in the sinus.
- *Aspergilloma*—it is mass of fungal hyphae seen in maxillary sinus.
- *Disseminated form*—this type of aspergillosis infection occur in case of immuno-compromised patient. Patient may have chest pain, cough and fever in this case. Prognosis of the patient is poor in this case.

Oral Manifestation

- *Sites*—it is seen on palate and tongue. Soft palate involvement seems to be more common in upper respiratory tract involvement.
- *Palatal lesion*—a lesion on the palate is manifested as a painful ulcer, surrounded by a ring of black necrotic tissue.
- *Gingival ulceration*—this is also a common feature of aspergillosis infection in oral cavity. The color of ulceration may be gray or violaceous hue.
- *Antroliths*—this is also reported from patient suffering from aspergillosis.
- *Disseminated infection*—oropharyngeal aspergillosis, in patients with hematological malignancies, presents as yellowish-black ulceration of soft palate and posterior part of tongue. These patients complain of intense local pain, oral bleeding and dysphagia.

Diagnosis

- *Clinical diagnosis*—gray color ulceration in oral cavity accompanied by nasal infection, chest pain may give clue to the diagnosis.
- *Laboratory diagnosis*—organism can be seen on biopsy.

Management

- *Amphotericin B*—it is the treatment of choice. It can be given intravenously.
- *Surgical debridement*—disseminated aspergillosis in immunocompromised patients should be treated on an individual basis surgical debridement.
- *Itraconazole*—itraconazole can be given local debridement of the lesion.

Protozoal Infection

Leishmaniasis

It is caused by the genus *Leishmania* which consist of three flagellate protozoa, which cause variety of distinct infections in man and are transmitted by sandfly bites.

Types

- *Visceral leishmaniasis*—it is also called as *Kalaazar*. It is caused by *Leishmania donovani*.
- *Cutaneous leishmaniasis*—it is caused by *Leishmania braziliensis*.

Clinical Features

Visceral leishmaniasis

- *Incubation period*—incubation period is 2 weeks to 2 years.
- *Symptoms*—onset may be insidious with a low grade fever or it may be abrupt with sweating and high intermittent fever. Cough and diarrhea can also develop.
- *Signs*—the spleen becomes enlarged, often massively. If not treated, patient will become anemic and wasted.

Mucocutaneous leishmaniasis

- *Incubation period*—is 1 week to 1 month.
- *Age*—it is usually seen in young men.
- *History*—there is past history of superficial ulcer of skin, caused by bite of an infected sandfly, which heals with depressed scar.
- *Nose*—nasal mucosa becomes congested and ulcerates. Later, all the soft tissues of nose may be destroyed.

Oral Manifestation

Visceral leishmaniasis

- *Pigmentation of face*—there may be increased pigmentation of face.
- *Gingival finding*—there may be spontaneous bleeding, edematous gingiva and loose teeth.

Mucocutaneous leishmaniasis

- *Incidence*—mucosal lesion usually occurs 1-2 years after skin lesion.
- *Site*—lips, soft palate and larynx may be involved.
- *Appearance*—the mucosal lesions are long standing, destructive, granulating ulcers which in many instance cause severe mutilation of structure involved.
- *Lymph nodes*—regional lymphadenopathy is common.

Diagnosis

- *Clinical diagnosis*—face pigmentation with edematous gingiva will suspect the disease.
- *Laboratory diagnosis*—organisms can be isolated from the lesion.

Management

- *Amphotericin B*—it is the drug of choice for visceral leishmaniasis. If the lesions are multiple, parenteral injection of amphotericin B should be given.

- **Cryosurgery**—in mucocutaneous lesion, small lesion may be treated by freezing with liquid carbon dioxide, curettage or infiltration with 1-2 ml sodium gluconate.

Trichinosis

It is caused by *Trichinella spiralis*, which is small, spiral, thread-like organism. Human infection occurs as a result of eating parasitized food, usually pork, which has not been completely cooked.

Clinical Features

- **Sites**—striated muscle, masseter, neck muscle and diaphragm.
- **Symptoms**—there is fever, facial and periorbital edema, muscle pain and eosinophilia.

Oral Manifestations

- **Site**—tongue is the most common site involved. It also occurs in muscles attached to mandible, in mandibular alveolar process and in gingival tissues.
- **Symptoms**—there is trismus, muscular cramps of the facial muscle, jaw and tongue. There is also monotony of speech.
- **Signs**—there may be petechiae of buccal mucosa, palate and floor of mouth. There is also bleeding from gingiva, lips and nose.

Diagnosis

- **Clinical diagnosis**—trismus, monotonous speech with petechiae of buccal mucosa or palate may give clue to the diagnosis.

Management

There is no specific treatment for trichinosis and in severe cases, prognosis is poor.

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Specific System Disorders

Basic Life Support

If the patient is unconscious you have to give him a basic life support. Patient may become unconscious in the dental chair and in that situation; you have to manage the patient in your dental clinics.

When victim heart ceases its activity, respiratory failure and cardiac arrest ensue. In such cases, administration of CPR within 3 minutes of cessation of activity of heart becomes a must.

Basic life support is the attempt to restore lung and heart function. Mainly, you have to follow the ABC of basic life support, i.e. **Airway, Breathing and Circulation**.

Positioning of the Patient

- *Supine position*—patient should be placed in supine position with the brain at the same level as the heart and the feet elevated slightly at 10 to 15 degree angulation. Head down position should be avoided as of gravity will act to force the abdominal viscera superiorly into the diaphragm, thus restricting movements.
- *Position of pregnant women*—if pregnant women loose consciousness in the dental office, the back of dental chair should be lowered to supine position and patient turn towards her right side, with a blanket or pillow under her back on her left to maintain that position.

Airway

Opening the airway and restoration of breathing constitutes the basic steps in life support. Any type of head support on the dental chair should be removed because such support flex the neck and thus making airway maintenance more difficult.

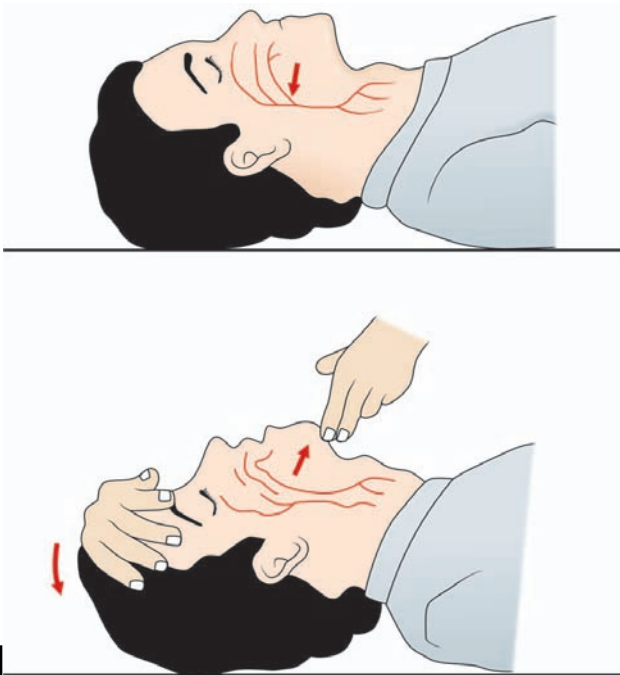
- *Head tilt*—this procedure is accomplished by placing the rescuer's hand on the victim's forehead and applying a firm, backward pressure with the palm.

- *Head tilt chin lift (Figs 33.1 A and B)*—in this, the fingers, of one hand are placed under the body- symphysis region of the mandible to lift the tip of the mandible up, bringing the chin forward. As the tongue is attached to the mandible, it is thereby pulled forward and off from the posterior pharyngeal wall.
- *Head tilt, neck lift*—in this, one hand of the rescuer is placed beneath the victim's neck to lift and support it.
- *Foreign material in airway*—if there is evidence of foreign matter in the airway, then check for airway patency. Partial airway obstruction produces noise and complete airway obstruction produces silence. If foreign body is suspected, the patient must be rolled on one side with lowering its head below the level of heart. Rescuer should place two fingers in the patient's mouth and remove anything in the entire oral cavity.

Breathing

You have to position your cheek close to victim's nose and mouth, look towards victim's chest; then look, listen and feel for breathing (5-10 second) (**Fig. 33-2**). Exhaled air ventilation can be give by mouth to mouth or mouth to nose.

- *Mouth to mouth*—if not breathing, pinch victim's nose close and give 2 full breaths into victim's mouth. This is done by taking a deep inspiration and exhaling in the patient's mouth and then patient is allowed to exhale passively. This procedure is continued at the rate of 12-14/min.
- *Mouth to nose*—when it is impossible to breathe into patient's mouth and if the rescuer if unable to adequately seal the mouth, the above technique is followed. In this technique, the rescuer keeps the head tilted backwards, with one hand on the forehead and other hand lifts victim's mandible, sealing the lips. Taking a deep breath, rescuer seals his lips around the victim's nose and blows in, until he feels and visualize the expansion of victim's lungs.



Figs 33-1A and B: Head tilt chin lift method to maintain the airway of the patient.



Fig. 33-2: Patency of air way should be checked.

- *Postbreathing*—after it, atmospheric air ventilation should be delivered to the victim. If breath does not go in, reposition the head and try again to give breath. If still blocked perform abdominal thrust (Fig. 33-3).



Fig. 33-3: Abdominal thrust should be given to patient if patient is not able to breathe.

Circulation

- *Carotid pulse checking*—you have to check for carotid pulse for 5-10 seconds, at side of victims neck
- *Rescue breathing*—if there is a pulse but victim is not breathing, give rescue breathing at the rate of 1 breath every 5 second or 12 breaths per minute
- *Technique to give compression to chest (Fig. 33-4)*—place heel of one hand on lower part of victim's sternum; with

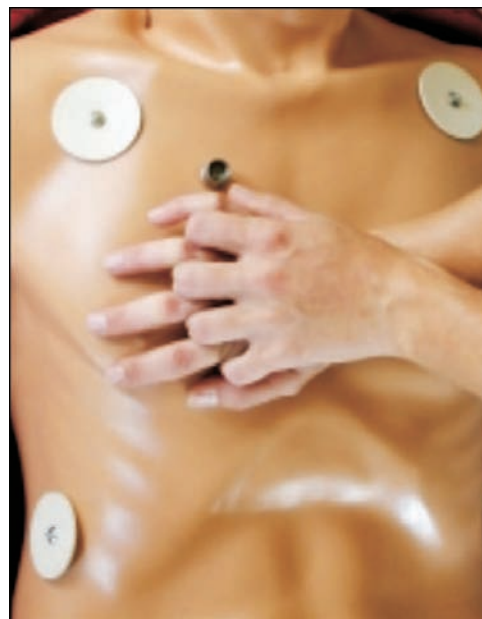


Fig. 33-4: Technique of describing chest compression.

your other hand directly on tip of first hand, depress sternum 1.5 to 2 inches. Perform 20 compressions to every 4 breaths (rate 80-100 per minute); check for return of pulse every minute. Continue it uninterrupted until advanced life support is available.

Cardiovascular Disease

Symptoms

- *Dyspnea*—it refers to breathlessness on physical exertion.
- *Orthopnea*—it is difficulty in breathing while lying down. It is due to rise in pressure in the right atrium and right ventricle leading to more blood flow into the lungs. Stiffness and congestion of lungs lead to breathlessness.
- *Edema*—edema of the feet and ankles is suggestive of cardiac failure due to retention of salt and water.
- *Pain*—causes of pain in chest are cardiac ischemia and pericarditis.
- *Palpitation*—it refers to awareness of the heart beat. It is a feature of anxious patient, paroxysmal tachycardia and atrial fibrillation.
- *Syncope*—it occurs due to decrease in cardiac output, decreased peripheral resistance or combination of the two.

Signs

- *Anemia*—pallor of skin and mucous membrane
- *Obesity*—such patients are more likely to have cardiac disease.
- *Peripheral cyanosis*—it occurs due to impairment of circulation, as in vasoconstriction, low cardiac output or stress.
- *Central cyanosis*—it occurs due to improper oxygen saturation of blood or due to right to left shunt.
- *Clubbing*—it is seen in congenital heart disease and in advanced cases of infective endocarditis (**Fig. 33-5**).
- *Temperature*—skin may be cold in cardiac failure and syncope.
- *Arterial pulse*—changes in the pulse pattern are noted in arrhythmias, reduced cardiac output, pulmonary embolism and aortic stenosis.

Examination of the Heart

- *Inspection*—enlargement of heart can be seen as a prominence to the left of sternum.
- *Palpation*—apex beat; normally apex beat is palpated within the mid-clavicular line in the 5th intercostal space. Displacement is seen in cardiac enlargement and fibrosis, collapse or removal of lungs.



Fig. 33-5: Clubbing in cardiovascular disease (Courtesy Dr Milind Chandurkar).

- *Dullness*—it is noted in pericardial effusion and emphysema.
- *Auscultation*—it is done to hear the heart sounds and murmurs.
- *First heart sound*—it is heard loudly at the apex. It occurs due to closure of the mitral valve. Accentuation of 1st heart sound is heard in anxiety and thyrotoxicosis, due to tachycardia and in mitral stenosis. Diminished heart sound is heard in myocarditis, cardiomyopathy and mitral regurgitation.
- *Second heart sound*—it is heard due to closure of the aortic and pulmonary valves. Second heart sound is altered in bundle branch block or increased ventricular stroke volume and from left or right shunt.
- *Third heart sound*—it is heard at the apex in young individuals. In older individuals, it is suggestive of mitral regurgitation and cardiomyopathy.
- *Fourth heart sound*—occurs due to forceful left ventricular distension before the first heart sound. It is suggestive of hypertension, cardiomyopathy, ischemic heart disease and left ventricular hypertrophy.
- *Murmur*—it occurs due to turbulent blood flow. It is heard due to abnormal heart valves.
- *Blood pressure*—it increases due to stress or tachycardia.

Consideration of Prophylaxis in Cardiovascular Disease

- *Heart disease in which prophylaxis recommended*—the disease in which prophylaxis recommended are prosthetic cardiac valve including bioprosthetic and homograft valve, previous bacterial endocarditis, surgically constructed systemic pulmonary shunt, rheumatic and other acquired valvular dysfunction, even after valve surgery, cardiomyopathy, mitral valves

prolapse with valvular regurgitation and most congenital heart malformations.

- *Heart disease in which prophylaxis is not recommended*—the disease in which prophylaxis is not recommended are surgical repair without residual symptoms beyond 6 months of atrial septal defect, ventricular septal defect, patent ductus arteriosus, previous coronary arterial bypass graft surgery, mitral valve prolapse without valvular regurgitation, heart murmurs, previous rheumatoid fever without valvular dysfunction and cardiac pacemaker and implanted defibrillation.
- *Dental procedure in which prophylaxis is recommended*—dental procedures likely to induce gingival or mucosal bleeding, including professional cleaning, surgical operations involving respiratory mucosa (including maxillary sinus), incision and drainage of infected tissues and intraligamentary injection.
- *Dental procedure in which prophylaxis is not recommended*—dental procedures not likely to induce gingival bleeding, injection of local anesthesia, shedding of primary teeth and new denture insertion will not require dental prophylaxis.

Angina Pectoris

It is hypoxia of the cardiac muscles, resulting from an imbalance between the oxygen consumption and oxygen supply of the cardiac muscles. It is the name given to paroxysm of pain. If anginal pain persists for more than half an hour, myocardial infarction or some acute abdominal condition should be considered.

Etiology

- *Coronary artery disease*—it is commonly associated with coronary artery disease and coronary artery spasm associated with atherosclerosis.
- *Oxygen carrying capacity of blood*—limited oxygen carrying capacity of blood and excessive oxygen demand.
- *Predisposing factors*—stress, physical or emotional or use of tobacco may predispose to an attack. Stress cause release of catecholamine and tachycardia.

Types

- *Stable*—it occurs with known physical effort and is relieved at rest and on administration of nitrates. It also gets aggravated by cold weather, smoking, emotional upset, high altitude, sexual excitement and straining at stools.
- *Nocturnal*—angina appears in the middle of the night due to left ventricular failure, which may be precipitate by dreams, causing release of catecholamine.

- *Unstable*—this is also called as peri-infarction angina as 20% of these patients develop myocardial infarction within 4 months. The following types of angina are called as unstable angina.
 - Recent angina (less than 60 days).
 - Stable angina in whom symptom are more severe in intensity, frequency and duration.
 - Angina at rest.
 - Angina following myocardial infarction.
- *Prinzmetal's angina*—this occurs in early morning, associated with ST segment elevation on ECG. It responds to nifedipine and nitrates as it is caused by coronary spasm which can be induced by smoking or hyperventilation.
- *Postinfarction angina*—some patients with myocardial infarction develop angina, 2 days to 8 weeks following the infarction.

Clinical Features

- *Age and sex*—it is most common in age range from 45 to 65 years and male to female ratio is 4:1.
- *Symptoms*—there is substernal pain radiating to both arms and ulnar border of the left arm, jaw, teeth, occipital region or epigastrium. The nature of pain is of crushing type. Pain is of short duration, lasting for 3 to 5 minutes. Pressure or discomfort in the substernal region. Patient may feel as if heavy weight has been placed on his chest. There may be breathlessness or fatigue, due to low cardiac output.
- *Relieving of pain after cessation of exertion*—in most of the cases, the pain is relieved by cessation of exertion. For this reason and because of intense pain, the person commonly maintains a fixed position during the attack.
- *Signs*—variation in pulse rate with generalized facial or circumoral pallor with cold perspiration may be exhibited.

Diagnosis

- *Clinical diagnosis*—a typical history of angina itself could give the diagnosis, even in absence of any other abnormalities on investigation or examination. Though physical examination in angina is often normal, certain clues to the presence of IHD may be present like gallop rhythm (third heart sound), left ventricular enlargement, thickened blood vessels and absent pulse, systolic murmur of mitral regurgitation or papillary muscle dysfunction.
- *Diagnostic tests*
 - *ECG*—in 50% of patients resting ECG may be normal.
 - *Stress testing*—with bicycle ergometer using standard protocols, or treadmill.

- *Thallium stress test*—done by injecting thallium, while the patient exercises and then regional myocardial perfusion is assessed by gamma camera.
- *Echocardiography and Doppler study*—this will be helpful to judge the regional wall motion, abnormality and left ventricular thrombus.
- *Coronary angiograms*—to diagnose blockade of coronary arteries and its location and severity.

Prevention of Anginal Attack

- *Modification of life style*—avoid exertion and walking uphill is avoided.
- *Tranquilizer*—diazepam 5-10 mg 6-8 hrly, to relieve anxiety.
- *Coronary vasodilator*—nitrates and calcium antagonist.
- *Control risk factor*—obese patients to reduce weight, smoking strictly forbidden.
- *Antiplatelet drugs*—low dose of aspirin should be given to prevent risk.

Management

- *Nitroglycerine tablet*—if the patient experiences an anginal attack in the dental chair, nitroglycerine tablet or sublingual spray (amyl nitrite) should be immediately applied under the tongue. Dose should be 0.5 mg of glycerine tri-nitrate or 5 mg of isosorbide dinitrate.
- *Prophylactic drugs*—in a known case of angina pectoris relatively short acting antianginal drugs such as sublingual isosorbide dinitrate tablet is recommended prophylactically, before initiating the dental therapy or a particularly stressful phase of dental therapy.

Dental Considerations

- *Difference between anginal pain and pain of dental origin*—the pain is usually referred to jaws and teeth, resembling a toothache and causing patient to seek dental attentions. Due to overlapping of the fifth cranial nerve, cardiac pain may be transmitted to jaws and interpreted as dental pain. Anginal jaw pain is characterized by its severity, its onset associated with exertion and its disappearance with rest. These characters differentiate it from the usual pain of dental origin.
- *Anginal attacks in dental chair*—acute anginal attacks may occur as a result of stress associated with dental services, i.e. extraction. If a patient is known to be angina pectoris patient, short acting antianginal drug such as sublingual isosorbide dinitrate is given prophylactically in addition to any long acting nitrate drugs before initiating dental therapy.
- *Deferring the procedure which require general anesthesia or conscious sedation*—any dental procedure which require general anesthesia or conscious sedation should defer for 3 months duration after recent anginal attacks.

Myocardial Infarction

It results from thrombotic occlusion (or sometimes prolonged spasm) of the infarct related blood vessels. Myocardial ischemia and necrosis occurs from subendocardial to subpericardial region. The entire process takes 6 hours to complete. An anginal attack lasting longer than 30 minutes is considered by definition to be myocardial infarction.

Clinical Features

- *Symptoms*—nausea, vomiting, tachycardia, grossly irregular pulse and breathlessness.
- *Signs*—there is pallor, diaphoresis and pulmonary edema. Initially, there is rise in blood pressure which may be followed by a fall, especially if cardiogenic shock occurs. Heart sounds are muffled. On second and third days, mild fever of 38-39° C may occur.

Diagnosis

- *History*—the characteristic history of chest pain and pain radiating to other areas, as described for angina may be the only guide. Patient who presents the history must be carefully evaluated because in early disease history may be the only positive finding.
- *Clinical symptoms*—clinical symptoms of collapse, severe unremitting chest pain, changes in heart rate, hypotension will give clue to diagnosis.
- *ECG changes*—the electrocardiogram shows changes characteristic of or compatible with acute myocardial infarction in only 80% of the cases and therefore cannot be used, always, to exclude the presence of myocardial infarction. Also shows the earliest change in ST segment elevation with the onset of pain.
- *Nonspecific test*—polymorphonuclear leucocytosis with high ESR may be seen in the first week, due to tissue necrosis.
- *Serum enzymes*—there may be elevation in certain enzymes like CPK, SGOT, LDH and AST in the blood.
- *Radionuclide technique* provides accurate information about myocardial infarction. In these, gammas emitting radionuclide are used in making cardiac scintigrams. Normally, perfuse myocardium is labeled by these radionuclide and abnormally perfuse myocardium is not labeled, producing the cold spot on scintigrams.
- *Positron emission tomography* is also very useful nowadays.

Management

- *Cardiopulmonary resuscitation*—in case of cardiopulmonary arrest, cardiopulmonary resuscitation should be immediately begun.
- *Prophylactic lidocaine*—in patients with life threatening acute arrhythmias, usually premature ventricular

contraction or ventricular tachycardia, occur in the dental office. Before transportation of the patient to an acute care medical facility, prophylactic 10% lidocaine can be injected IM in the deltoid muscle at the dose of 4 to 6 mg/kg body weight.

- Control of chest pain is done by *sublingual nitroglycerine* 0.3-0.4 mg, repeated every 5-10 minutes and morphine hydrochloride 15 mg, subcutaneously.
- *Nitrous oxide* in a concentration of 35%, mixed with oxygen has been quite effective in decreasing the pain of acute myocardial infarction.
- *Anticoagulants* like heparin 2000-5000 units, every 6-8 hourly.
- *Oral aspirin*—oral administration of aspirin daily improves survival but nowadays it is not recommended for daily use.

Dental Consideration

- *Premedication*—these patients require special management because they may have decreased ability to withstand stressful conditions. Patients, for whom dental treatment is particularly stressful, should be premedicated before treatment. Oral benzodiazepam are helpful in 5 mg doses. It should be given in a waiting room 45 minute before the procedure.
- *Patient on anticoagulant therapy*—careful history taking is a must in such patients. Patients on long-term anticoagulant therapy should be given instructions with regard to bleeding, if surgical procedures like dental extraction are carried out. Anticoagulant dosage should be decreased by the cardiologist before any surgery. Sudden withdrawal of anticoagulant drugs may result in thrombosis or embolism.
- *Timing of dental treatment*—after acute myocardial infarction all dental procedure should be deferred for 3 weeks and after that simple emergency dental treatment can be carried out but with the opinion from physician.

Rheumatic Heart Disease and Fever

It is an inflammatory complication that may follow group of streptococcal infection; manifested by one or more of the following arthritis, carditis, and chorea. Rheumatic fever usually comes on 1 to 3 weeks after the streptococcal infection.

Clinical Features

- *Age*—it is usually a disease of childhood occurring most often between the ages of 6 to 16 years with peak of 8 years.
- *Symptoms*—chorea (involuntary movement), the symptoms of acute carditis. The child often complains of sore throat and has a temperature of 38 to 39°C.

- *Signs*—typical subcutaneous nodules may be present. There is involvement of successive joint which are red, tender and painful. Wrist, ankles, elbows and knees are commonly involved (**Figs 33-6A and B**). Murmur and erythema marginatum, epistaxis and abdominal pain.



Figs 33-6A and B: (A) Hand deformity in rheumatoid arthritis (B) same hand in palmar view (Courtesy Dr Milind Chandurkar).

Diagnosis

- *History*—history of preceding streptococcal infection and predominate manifestation of arthritis and carditis.
- *Jones diagnostic criteria*—Jones criteria of diagnosis of rheumatic fever. At least of two major criteria should be followed.

Major criteria	Minor criteria
• Carditis • Polyarthritis • Chorea • Subcutaneous nodule • Erythema marginatum • Characteristic ECG change	• Fever • Arthralgia • Previous rheumatic fever or rheumatic heart disease • Elevated ESR and positive C-reactive protein • Protein raised

Management

- *Rest*—bed rest and sedation during acute episode.
- *Salicylates*—salicylates are almost specific for the pain of rheumatic fever.
- *Prophylaxis*—continued prophylaxis against a beta hemolytic streptococcal infection is indicated and given in the form of a monthly injection of 1.2 million units of benzathine penicillin G or in 200,000 units of oral penicillin, given twice daily or 1 g of sulfadiazine, given orally, once a day. Such prophylactic therapy is discontinued after 20 to 39 years of age.

Dental Considerations

Following are suggestions for prophylactic procedures, for reduction of postextraction bacteremia and the possible development of subacute bacterial endocarditis -

- *History*—proper history taking about evidence of rheumatic fever, MV prolapse. If there is history of rheumatic fever, administer prophylactic antibiotic therapy before dental procedure.
- *Antibacterial mouth rinse*—make the patient rinse with antibacterial mouthwash, such as chlorhexidine immediately before the treatment to help reduce the number of microorganisms.
- *Atraumatic procedure*—keep dental procedure atraumatic as best possible.
- *Refer to physician*—if a febrile illness develops within 3 months of the dental procedure, refer the patient to physician.

Hypertension

Hypertension is the most common and significant medical condition encountered in dental practice. Normal blood pressure is 120/80 mm Hg.

Stages

- *Stage I (mild) hypertension*—a diastolic pressure of 90-99 mm Hg and systolic pressure of 140-160 mm Hg.
- *Stage II (moderate) hypertension*—diastolic pressure of 100 to 109 mm Hg and systolic pressure 160-170 mm Hg.
- *Stage III (severe) hypertension*—it has diastolic pressure 110-119 mm Hg and systolic pressure of 170-190 mm Hg.
- *Stage IV (very severe) hypertension*—it is also called as *malignant hypertension*. It has diastolic pressure more than 120 mm Hg and systolic pressure of 190 mm Hg.

Types

- *Primary or essential or idiopathic hypertension*—it occurs due to hereditary or environmental cause. Environmental cause includes high sodium intake, excessive alcohol consumption, physical inactivity, smoking and stress.

- *Secondary hypertension*—it may be caused by factors such as renal parenchymal disease, renal artery disease, pheochromocytomas, adrenal cortical hyperfunction and central nervous system lesion.

Clinical Features

- *Symptoms*—patient may complaint of occipital headache, dizziness, nausea, vomiting, malaise, and shortness of breath and nose bleed. In some cases chest pain occurs. In a milder form, there is extensive intraoperative bleeding.
- *Signs*—blood pressure above 160 / 100 mm Hg. There is cardiac enlargement and narrowing of retinal arterioles also occurs. Hypertensive patients may lead to hemorrhage, myocardial infarction, cardiac decompression and renal failure.
- *Odontalgia*—odontalgia is observed in hypertensive patients, which occurs due to hyperemia of dental pulp or congestion of this tissue resulting from increase blood pressure.
- *Drug side effect*—many patients in dental office may be reported with gingival enlargement which occurs due to nifedipine (calcium channels blocker) use in hypertensive patients. Antihypertensive medication like thiazides, methyldopa, propranolol, etc. can cause lichenoid reaction in the oral cavity. ACE (angiotensin converting enzyme) inhibitors can cause taste impairment, burning sensation of mouth called as *scalded mouth syndrome*. Xerostomia can also occur due to many antihypertensive drugs.
- *Hypertensive crisis*—hypertensive crisis is a sudden rise of arterial blood pressure to a very high level (160/100 or 250/150 mm Hg) with clinical and pathological manifestations. Such patients can pose problems involving heart, brain and kidney.

Dental Significance

- *Treatment planning*—an elevated blood pressure in a dental patient requires careful consideration in treatment planning, premedication, selection of an anesthetic and determining the duration and extent of operative procedure.
- *Risk of medical emergencies*—undetected hypertension increases the risk of experiencing cardiovascular conditions such as angina, myocardial infarction and cerebrovascular accident while undergoing dental care.
- *Effects of antihypertensive drugs*—oral health care can be affected by the known side effects of antihypertensive drugs and their interaction with other medications.
- *Care of general health*—the dentist can perform a valuable patient service by identifying undetected hypertension.

Dental Management

- *Identification*—the dentist should have equipments in the office to measure blood pressure.
- *Pain control*—effective control of operative and post-operative pain should be done after dental procedure.
- *Stress and anxiety reduction*—elevation of blood pressure caused by nervousness or stressful situations should be minimized in hypertensive patients. The patient should not be permitted to struggle when using inhalation analgesia or general anesthesia and adequate oral and parenteral premedation should be used. Patients with known hypertension should have blood pressure checked at each dental visit, to assure that there is no risk of harm from the stress of dental procedure. Very anxious patient should be given 5 mg of diazepam or 30 mg of oxazepam, night before and one hour before the dental appointment.
- *Limit the use of epinephrine*—epinephrine use should be limited in patient with hypertension. The reason for this many patients of hypertension usually suffer from the cardiovascular disease and epinephrine will cause vasoconstriction in these patients.
- *Avoidance of drug interaction*—many NSAID interacts with ACE inhibitors decreasing the efficacy of antihypertensive agents. Vasopressor drugs can interact with non-selective beta blockers provoking hypertensive episode.
- *Physician referral*—if blood pressure is high, patient should be referred to the physician for further observation and regarding the antihypertensive medication.
- *Emergency extraction*—when extraction is necessary on an emergency basis, the hypertensive patient should be hospitalized and local homeostatic measures should be taken to avoid undue hemorrhage. In these patients, proper premedication should be given.
- *Hypertensive crisis in dental chair*—if it arises intra-operatively, patient should rest in semisitting position. O₂ may be administered; if it does not help, diazepam may be administered at very slow rate. If this is not helpful, the patient should be left to the care of medical physician. In the mean time, injection frusemide is given intravenous and 10 ml capsule of nifedipine may be broken and sprinkled sublingually to bring the blood pressure down.
- *Management of side effects of drugs*—xerostomia should be managed by pilocarpine, sugarless mint or gums and minimizing caffeine intake. Gingival overgrowth is reduced by excellent oral hygiene. Lichenoid reaction is treated by topical corticosteroids.

Infective Bacterial Endocarditis

It is a serious disorder which is the most common bacterial origin but on occasion, it may be mycotic. Causes of

endocarditis of dental origin are almost always caused by bacteria of low virulence, that slowly attack a previously damaged endocardium, causing subacute bacterial endocarditis.

Pathogenesis

- *Organism responsible*—it is usually caused by oral microorganism like alpha hemolytic streptococci, enterococci, pneumococci, staphylococci and group A streptococci.
- *Damaged blood flow*—in normal patients blood becomes sterile after this transient phase. In patients with damaged heart valves, initially there is formation of a sterile platelet fibrin clot or thrombus on the damaged heart surface. It acts as nidus for bacterial proliferation, subsequently leading to infective endocarditis.
- *Oral bacteremia*—dental procedure can lead to release of bacteria into the blood circulation. The most common type of organism responsible for this oral bacteremia is streptococci viridians which are abundantly found in the mouth. It occurs in person who is having poor oral hygiene and undergoing extraction.
- *Consequence of infective endocarditis*—infective endocarditis will lead to proliferation of bacteria on the damaged heart surface can lead to impaired valvular function, congestive heart failure, abscess of myocardium due to production of pus and release of infectious emboli which can get lodged in organs such as brain, kidney and spleen.

Predisposing Factors

- *Heart disease*—it has marked predisposition for persons with rheumatic or congenital, cardiac or vascular defect, bicuspid aortic valve, leutic aortic valvular disease, idiopathic hypertrophic subaortic stenosis and mitral valve prolapse.
- *Surgical correction*—recent surgical correction of congenital valvular defect within 6 months.
- *Hypertrophic cardiomyopathy*—it can also occur in hypertrophic cardiomyopathy.
- *Surgical trauma*—surgical trauma and dental extraction are commonly related chronologically, to the onset of clinical symptoms.

Clinical Features

- *Age*—it can occur at any age, but most common in middle age group.
- *Progress*—after formation of vegetative lesion (thrombi), they serve as foci of the intermittent dissemination of microorganisms throughout the body. These vegetative lesions are friable and small pieces may break off and form septic emboli.

- *Symptoms*—the patients experience progressive weakness, loss of weight, dyspnea, anorexia, muscular and joints aches and pains, with low grade fever.
- *Signs*—Petechial hemorrhage in the conjunctiva and oral mucosa occur due to septic emboli.
- Larger emboli can lodge occasionally in spleen, kidney, lungs and brain where they produce symptoms referable to the structure and the organs involved.

Diagnosis

- *Clinical diagnosis*—it is frequently made by elimination of other conditions. The disease should be suspected in any patient with valvular heart disease, who has unexplained fever for week or more, or exhibits embolic phenomenon or an unexplained anemia.

Management

- *Antibiotics therapy*—intensive antibiotic therapy, usually 20,000,000 units of penicillin in combination with gentamicin for 2 weeks should be given.
- *Removal of infected valve*—early removal of infected valve should be done with sterile replacement.

Dental Considerations

- *Oral cavity, a source of infection*—organisms responsible for subacute bacterial endocarditis were disseminated commonly through bloodstream, following dental extraction. It was demonstrated that gingival sulcus was an important site from which the bacteria gained entrance into the bloodstream. Actual cauterization of gingival crevice before extraction markedly reduces the percentage of transient bacteremia. Even in the absence of direct dental procedure, bacteremia can be produced secondary to foci of infection in the oral cavity.
- *Prophylactic measure*—every known prophylactic measure should be taken to preclude the possibility of transient bacteremia occurring in patients with known vascular lesions.

Prevention

- *Proper history should be taken*—ask the patient about history of rheumatic fever, heart disease with valvular involvement or heart murmur.
- *Prophylactic antibiotics therapy*—if the heart disease to endocarditis is known to be present, administer the prophylactic antibiotic therapy before the dental treatment.
- *Physician consultation*—if questionable positive history of heart disease is obtained, you have to consult the patient's physician. All patients who are at risk of developing endocarditis, subsequent to dental treatment,

should be instructed to consult physician if a febrile illness develops within 3 months of a dental treatment.

- *Antibacterial mouth rinse*—make the patient rinse with an antibacterial mouthwash, immediately before dental treatment, to help reduce the number of oral micro-organisms.
- *Atraumatic dental procedure*—keep dental treatment as atraumatic as possible.

Congestive Cardiac Failure

It is the condition where the heart fails to maintain an output sufficient for the needs of the body.

Classification

- *Low output cardiac failure*—there is a primary lesion in heart which decreases the contractibility of the heart and causes diminished cardiac output.
- *High output cardiac failure*—there is primarily no lesion in the heart, but due to extra cardiac condition, there is increased work load on the heart which causes cardiac failure with increased cardiac output.

Causes

- *Low output failure*—it is caused by myocardial lesions like ischemic heart disease, rheumatoid heart disease, cardiomyopathy, valvular endocarditis, congenital heart disease and vascular lesions like hypertension and aneurysm of aorta.
- *High output failure*—thyrotoxicosis, anemia, hypoproteinemia, beriberi, AV Fistula and cirrhosis of liver can cause congestive cardiac failure.

Clinical Features

- *Symptoms*—there is increased breathlessness following moderate exertion, chronic productive cough associated with blood tinged sputum. Patient also complains of wheezing, anorexia, bronchospasm, and dyspnea.
- *Signs*—pitting edema of lower extremities, hepatic enlargement, and generalized edema. Congestion of the large veins of neck and raised jugular venous pressure. There is also cyanosis of lips, tongue and oral mucosa with ankle edema.

Diagnosis

- *Clinical diagnosis*—breathlessness, generalized edema and raised jugular venous pressure will give clue to diagnosis.

Management

- *Bed rest*—complete bed rest is required.
- *Digoxin*—it should be the first line therapy for heart failure.
- *Diuretic*—in cardiac failure, there is always sodium and water retention, hence diuretics are given.

- *Vasodilators*—vasodilators are given, e.g. nitroprusside—5-10 mg/min.
- *Oxygen*—it can be administered for proper circulation.
- *Massage*—massage of lower limb to maintain peripheral circulation.

Dental Considerations

- *Heart attack in dental office*—the dentist should watch for early signs of congestive cardiac failure like cyanosis of lips, tongue. If the patient develops acute pulmonary edema in the dental office, patient should be made comfortable in sitting position or semisitting position and administered 100% O₂. Injection frusemide and aminophylline should be given. Physician should be immediately called for.

Congenital Heart Disease

Congenital heart diseases are the most common types of anomalies present in child and more common as compared to rheumatic diseases.

Clinical Features

- *Symptoms*—they are variable. Some children manifest breathlessness, inability to thrive, growth retardation and central cyanosis.
- *Persistent ductus arteriosus*—it is failure of closure of ductus arteriosus which plays role in fetal life as most of the blood passes through ductus arteriosus into the aorta. It results in recirculation of blood through the lungs, leading to increased workload on the heart. It occurs commonly in women. Affected individuals may suffer from growth retardation, dyspnea, cyanosis and cardiac failure. Surgical correction is possible in some cases.
- *Atrial septal defect*—it results in shunting of blood from left atrium and then to the right ventricle and pulmonary arteries, leading to progressive enlargement of the right side of the heart and the pulmonary artery with its branches. Females are affected more than males. There is dyspnea, cardiac failure and arrhythmias. It is treated by surgical closure.
- *Ventricular septal defect*—it is usually asymptomatic.
- *Pulmonary stenosis*—it occurs either alone or along with atrial septal defect or ventricular septal defect. Larger pulmonary stenosis results in dyspnea, fatigue or syncope.

Oral Manifestations

- *Bluish red discoloration*—general bluish red discoloration of oral mucosa with severe marginal gingivitis and bleeding.

- *Tongue*—tongue is often deeply fissured and edematous.
- *Delayed eruption*—both permanent and deciduous teeth are delayed in eruption.
- *Enamel hypoplasia*—this is also frequently present in patient with congenital heart disease.
- *Skimmed milk appearance*—teeth will have bluish white skimmed milk appearance with vasodilation in the pulps.

Diagnosis

- *Clinical diagnosis*—generalized bluish discoloration of oral muscle with symptoms of atrial septal defect and persistent ductus arteriosus.

Management

- *Antibiotics prophylaxis*—antibiotics cover is needed for operative procedure.
- *Cardiac surgery*—this should be done to correct congenital anomalies.

Dental Considerations

- *Adequate analgesia*—adequate analgesia should be given to the patient during oral surgical operation. General anesthesia should be avoided.
- *Conscious sedation*—conscious sedation with nitrous oxide should be given with approval of physician.
- *Avoid using gingival retraction cord containing epinephrine*—gingival retraction cord containing epinephrine should be avoided.

Cardiac Transplantation

These patients are on immunotherapy for life. Too little therapy will result in rejection of episode, while too much runs the risk of both infection and neoplasm. Opportunistic infections like fungal, protozoal and viral infection are common in these patients.

Dental treatment after transplantation should be carefully planned in view of the patient's lower immune status with particular attention to WBC count and Hb concentration. Before performing any treatment, the dentist should take consent of the patient's physician and consider antibiotic prophylaxis. As many of these patients are on long-term steroid therapy, steroid supplements should be given.

Prophylactic antibiotic dose and its indications:

- *Amoxicillin*—3 gm orally, 1 hr before the procedure. 1.5 gm 6 hours after the initial dose.
- *Patients with allergy to penicillin*—Erythromycin ethyl succinate 800 mg, Erythromycin stearate 1 gm orally, 2 hours before the procedure. Clindamycin 300 mg 1 hour before the procedure and 150 mg, 6 hours after the initial dose.

Coarctation of Aorta

It is a developmental anomaly characterized by marked diminution in the caliber of aortic arch, just distal to where the left subclavian artery arise.

Patient suffers from secondary hypertension and cerebral aneurysm with hemorrhage.

Intraorally there is marked enlargement of mandibular arteries and branches leading to individual teeth. Arterial hemorrhage will occur following tooth extraction. Pulp of all the four maxillary incisors were markedly enlarged and funnel shaped, occupying great portion of crown and roots.

Respiratory Disorders

Bronchial Asthma

Asthma is a common disease estimated to affect 4 to 5% population. It is initially a spontaneously, reversible, spasmodic contraction of the smooth muscles of bronchi resulting in bronchiolar narrowing. As the attack progresses, resistance to airflow may be further exacerbated by mucosal edema and inflammatory infiltrate of mucosa.

Types

- *Extrinsic asthma*—it is developed by allergic factors. It is also called as allergic asthma. Patient, who is having asthma of allergic origin, often has positive family history of disease and positive atopic history including hay fever, rose fever and eczema.
- *Intrinsic asthma*—it is precipitated by nonallergic factors. It is also called as *non-allergic asthma*, *idiopathic asthma* and *infective asthma*.
- *Mixed asthma*—it is combination of allergic and non-allergic asthma.
- *Status asthmaticus*—it is persistent exacerbation of asthma.

Predisposing Factors

Extrinsic asthma

- *Airborne allergen*—house dust, feathers, animal dander, furniture stuffing, fungal spores and a wide variety of plant pollens.
- *Allergenic food*—it includes cow's milk, eggs, fish, chocolate, shellfish and tomatoes.
- *Allergic drug*—it includes penicillin, vaccines and aspirin.

Intrinsic asthma

- *Infections*—respiratory infections, usually viral, are well known initiating factor in the asthmatic patient.
- *Exercise*—some patients have acute asthmatic attack after prolonged exercise.

- *Psychological*—emotional stress such as nervousness, anxiety can cause asthmatic attack. The dental office is a common site for asthmatic attack. Child may develop asthmatic attack after taken into treatment room and recover after moving out from treatment room.

Clinical Features

- *Age*—it is more common in children, especially boys. It can also occur in older individuals.
- *Symptoms*—patients notice a sensation of fullness in the chest. Predominately, symptoms of acute attack are wheezing, coughing and labored breathing. There is also sneezing and gasping sounds are heard while attempting to breath.
- *Attack termination*—termination of attack is usually heralded by a period of intense coughing with expectoration of thick, tenacious mucous plug which is followed by sensation of relief.
- *Signs*—with severe attack, the patient is extremely anxious and agitated. Heart rate is increased to 130 per minute. Cyanosis of mucous membrane of lips may be visible along with perspiration and flushing of face and upper torso in severe attack. Patient is more comfortable if allowed to sit or stand upright with the back upright and the chest, shoulder and head fixed.
- *Extrinsic asthma*—bronchospasm usually develop within minutes after exposure to allergens. These attacks usually become less frequent and less severe during middle and late adolescence and may disappear entirely.
- *Intrinsic asthma*—attack of intrinsic asthma is usually more fulminant and severe than those of allergic asthma. The long-term prognosis is poorer and the patient eventually exhibits clinical signs and symptoms in interval between acute episodes.
- *Status asthmaticus*—in it, acute asthmatic attack persists in spite of drug therapy. Bronchospasm may continue for hours or even days without remission. Patient more commonly exhibits extreme fatigue, dehydration, severe hypoxia, cyanosis, peripheral vascular shock and drug intoxication from intensive therapy. Chronic partial airway obstruction may lead to death from respiratory acidosis.

Diagnosis

- *Clinical diagnosis*—diagnosis of bronchial asthma is based on the symptoms, pulmonary function test and physical finding of expiratory wheezes during the acute attack.
- *Laboratory diagnosis*—there is raised total IgE and specific IgE antibody concentration.

Management

- *Sympathomimetic amine*—use of drugs like terbutaline, isoproterenol and metaproterenol as first line therapy. It dilates and prevents bronchial smooth muscle constriction.
- *Xanthine derivatives*—xanthine derivatives like aminophylline and theophylline can be used. It inhibits the hydrolytic degradation of cyclic AMP.
- *Corticosteroid*—corticosteroids like hydrocortisone and prednisone can also be used. It lessens intensity of antigen-antibody reaction.
- *Omaliuzumab*—it is recombinant humanized monoclonal anti-IgE antibody, will cause reduction in total serum IgE and thereby decreasing the symptoms of asthma.
- *Sodium cromoglycate*—cromolyn sodium is used. It protects against mast cell destruction.
- *Combination*—inhaled corticosteroids with bronchodilators.
- *Antihistamine*—antihistaminic usually are useful because they produce drying and obstruction of air.
- *Emergency management*—emergency treatment is inhalation of a solution containing 0.1 mg isoproterenol or 1:1000 epinephrines by nebulizer or injection of 0.1 ml of 1:1000 epinephrines. Continuous inhalation by nasal catheter is helpful to relieve the hypoxia.
- *Hydration*—hydration is necessary in the form of 5% glucose in water because there is dyspnea.

Dental Considerations

- *Avoid inhalation anesthetics*—patient using beta-adrenergic inhaler should be reminded to bring the inhaler with them to dental office; the dentist should avoid inhalation anesthetics or analgesic in asthmatic patient because of the possibility of stimulating an acute asthmatic attack.
- *Dental work*—if attacks are seasonal, routine dental work can be performed during the time when frequency of attack is lowest. Patients on steroids may require additional steroid to avoid serious reaction to stress of dental procedure.
- *Attacks in dental chair*—in case of acute attack occurring on dental chair, following measures should be taken:
 - *Terminate the procedure*—terminate the dental therapy and position the patient in any comfortable position.
 - *Administration of bronchodilators*—0.5 ml, 1:1000 adrenelines can be injected subcutaneously or intramuscularly. If attack is prolonged, steroids are indicated
 - *Aerosol spray*—the onset of aerosol drug is rapid and relief of symptoms occurs within seconds.
 - *Administer oxygen*—it may be administered by full face mask, nasalhood or nasal cannula.

Chronic Obstructive Pulmonary Disease

It is characterized by airway obstruction and breathlessness.

Types

- *Chronic bronchitis*—it refers to inflammation of bronchi. It is defined as a condition in which there is mucus producing cough present, for at least 3 months of the year, for more than 2 consecutive years.
- *Emphysema*—it is dilatation of air spaces distal to terminal bronchioles with destruction of alveoli reducing alveolar surface area for respiratory exchange.

Causes

- *Smoking*—the most common cause of COPD is smoking.
- *Environmental dust*—the other factors are chronic recurrent infection, air pollution and occupational inhalants.
- *Deficiency of antiproteolytic enzyme*—deficiency of anti-proteolytic enzyme alpha 1-antitrypsin is a rare cause of emphysema.

Clinical Features

- *Early morning cough*—patient shows early morning mucoid cough which becomes purulent during exacerbation.
- *Pink panter or pink puffer*—this is present in emphysema in hyperventilation, occurs to maintain normal blood gases. This will cause vasodilatation and patient will be pink due to CO₂ retention.
- *Cutaneous vasculitis*—in some cases of emphysema cutaneous vasculitis may be present (**Fig. 33-7**).



Fig. 33-7: Cutaneous vasculitis seen in respiratory disease (Courtesy Dr Milind Chandurkar).

- *Blue bloated appearance*—this is present in chronic bronchitis. In this patient is unable to maintain hyperventilation and become hypoxic. This will lead to central cyanosis, ankle edema, and raised jugular venous pressure of cor pulmonale to give blue bloated appearance. Wheezing, dyspnea and cough.
- *Later stage*—in the later stage, there can be blood tinged sputum and fever.

Diagnosis

- *Clinical diagnosis*—cutaneous vasculitis with pink panter with blue bloated appearance will aid in diagnosis.

Management

- *Stoppage of habit*—patient should ask to quit the habit of smoking.
- *Bronchodilator*—bronchodilators are particularly effective in relieving bronchospasm. Bronchodilators used are ipratropium bromide, oxitropium, and theophylline.
- *Antibiotics*—antibiotic therapy should be initiated with earliest sign of chest infection. Antibiotics used are amoxicillin, trimethoprim, or tetracycline.
- *Oxygen therapy*—oxygen is useful both, intermittently on a chronic basis and continuously with an acute exacerbation of disease.

Dental Considerations

- *Upright position*—patient should be treated in upright position. If patient is flat there are chances of breathlessness.
- *Avoid using rubber dam*—the patient's tolerance to partial airway obstructing devices such as rubber dam is less so use of rubber dam should be avoided.
- *Short treatment time*—mucus producing cough, wheezing, and dyspnea in these patients prevent long treatment sessions. This should be taken into consideration while planning the dental therapy.
- *Inhalation analgesic*—inhalation analgesic should be given only when it is necessary and in conjunction with anesthesiologist.
- *Drug interaction*—as patient is taking theophylline, many drugs like epinephrine, erythromycin, clindamycin, etc. should be given cautiously.
- *Avoid giving general anesthesia*—patient should be avoided giving general anesthesia unless it is very necessary.

Pulmonary Foreign Bodies

Foreign bodies occluding or obstructing a main respiratory passage will produce cyanosis and asphyxia. Sudden and

violent attacks of coughing, accompanied by shortness of breath and chest pain are classic symptoms of pulmonary foreign bodies.

Dental procedures are directly or indirectly involved in a high percentage of foreign body problems hence, the cooperation of the dentist is essential for the prevention of these accidents. Dentures are important indirect cause of foreign body accident. The tactile sense of the denture patient is severely impaired by the dentures and the foreign body (usually bones) may pass the point of recovery by reflex action before the patient becomes aware of it.

Dentures can become embedded in the soft tissue or block the respiratory and alimentary tract, without being detected by the X-ray (as denture material is radiolucent) such as in high velocity automobile accidents.

Denture foreign body can be a portion of tooth, a restoration or an operating instrument that is lost during dental treatment, tooth fragment, instrument during surgery and prosthesis lost by the patient.

The risk of respiratory tract foreign body can be eliminated almost entirely by the use of the rubber dam or throat packs. Complete and partial dentures should be removed during sleep, unconsciousness and before surgery.

Renal Disorder

Renal Failure

Renal failure produces a standard symptom complex, regardless of the underlying cause. It is caused by many diseases.

Etiology

- *Glomerulonephritis*—it represents a heterogeneous group of disease of varying etiologies that produce irreversible impairment of function. The attack of glomerulonephritis is either streptococcal or non-streptococcal. Chronic glomerulonephritis has usually very slowly, but steadily progressive course, leading to renal failure or uremia in few years to as many as 30 years.
- *Pyelonephritis*—it refers to the effect of bacterial infection in kidney, with *E. coli* being the cause of infection. Any obstruction of urinary tract can predispose to active pyelonephritis and also can occur due to generalized sepsis in patients with bacterial endocarditis or with staphylococcal infection. Clinically, there is sudden rise of body temperature, shaking chills, aching pain in one or both costovertebral areas or flanks and symptoms of bladder inflammation.
- *Hemolytic uremic condition*—this condition is caused by *E. coli*.

- *Other diseases*—other diseases which can cause renal failure are polycystic renal disease, nephrosclerosis, diabetic nephropathy, collagen vascular disease, hereditary nephropathy, analgesic abuse nephropathy, obstructive nephropathy, gouty nephropathy, neoplastic nephropathy, etc.

Clinical Features

- *Gastrointestinal*—nausea, vomiting, anorexia, parotitis, gastritis and gastrointestinal bleeding.
- *Neuromuscular*—headache, myoclonic jerks, peripheral neuropathy, paralysis, seizures and asterixis.
- *Hematological and immunological*—normocytic and normochromic anemia, coagulation defects, increased susceptibility to infections, decreased erythropoietin production and lymphocytopenia.
- *Endocrine and metabolic*—renal osteodystrophy, secondary hyperparathyroidism, impaired growth and development, loss of libido and sexual function and amenorrhea.
- *Cardiovascular*—arterial hypertension, congestive heart failure, cardiomyopathy, pericarditis and arrhythmias.
- *Dermatological*—pallor, hyperpigmentation, ecchymosis, uremic frost, pruritus, reddish brown distal nail beds.

Oral Manifestations

- *Incidence*—in the study of renal patients, 90% were found to have oral manifestations.
- *Ammonic taste and smell*—patients complain of *ammonic taste and smell*, particularly in the morning. It is caused by the high concentration of urea in saliva and its breakdown to ammonia.
- *Xerostomia*—the patients may complain of *xerostomia* which is caused by direct involvement of salivary gland, chemical inflammation, dehydration and mouth breathing.
- *Erythromopultaceous form*—the acute rise in BUN level may result in uremic stomatitis, which appears as *erythromopultaceous form*, characterized by red mucosa covered with a thick exudate and a pseudomembrane.
- *Ulcerative form*—in some cases, stomatitis may be in *ulcerative form* which appears as frank ulceration with red and pultaceous coat.
- *Low caries activity*—low caries activity despite of high sugar intake; poor oral hygiene due to increased salivary urea nitrogen.
- *Enamel hypoplasia*—it is frequently seen in patients where renal disease is started at young age.
- *Other*—there is pulpal narrowing and calcification, severe tooth erosion and loss of lamina dura can also be seen.

Diagnosis

- *Clinical diagnosis*—variety of clinical symptoms occur in it with oral manifestation.
- *Radiological diagnosis*—pulpal narrowing with loss of lamina dura can be seen.

Management

- *Hemodialysis*—to remove nitrogenous and toxic products of metabolism from blood by means of a dialysis system.
- *Peritoneal dialysis*—in it, 1-2 liters of dialysate are placed in the peritoneal cavity to remain there for varying intervals of time. Main advantage of this is that there is no risk of air embolism and blood leak.
- *Drug treatment*—Aminoglycoside (gentamicin), antimicrobial (penicillin), analgesic (acetaminophen), narcotic (codeine, morphine), sedative (diazepam), amitryptline, antihistaminic (chlorpheniramine), phenytoin and lidocaine.
- *Kidney transplantation*—it involves surgical removal of a kidney from a donor and implantation into a recipient. This method is for prolonged life in patient with end stage renal failure; the kidney transplantation patient receives a continuous regimen of immunosuppressive medication to ensure graft survival.

Dental Consideration

- Many patients who are receiving dialysis have conditions of oral neglect.
- *Dialysis*—if time and patient's conditions permit, dialysis should be part of the preoperative preparation. Dialysis will return the state of hydration, serum electrolyte, urea nitrogen and creatinine toward normal and will reduce the need for dialysis in the immediate postoperative period.
- *Control of bleeding*—as bleeding is common problems in renal failure patient, hemostasis should be done with the help of desmopressin, cryoprecipitate and conjugated estrogens.
- *Position of patient in dental treatment*—blood flow through the arm should not be impeded by requiring the patient to assume a cramped position or using that arm to measure blood pressure. Patient should avoid sitting for long periods of time with leg dependent. If dental treatment is long, then patient should be allowed to walk for few minutes every hour.
- *Infective endocarditis*—the presence of an access site increases the susceptibility to infective endocarditis. Antibiotic prophylaxis is required for control of infective endocarditis. Start with broad spectrum antibiotic, e.g. amoxicillin 3 g, 1 hr before treatment and 500 mg every 8 hours for 1-2 days.

- *Candidial infection*—candidal infection can occur in oral cavity. For candidiasis, administer nystatin mouthwash 5000000u/ml QID, day before the treatment and 2 days after dental treatment.
- *Medication in dialysis patient*—most drugs are excreted at least partially by the kidney. Following are the drugs to be avoided in dialysis patients:
- *Nonsteroidal anti-inflammatory drugs*—it may induce sodium retention and impair the action of diuretics, prevent aldosterone production and cause acidosis.
- *Tetracycline and steroids*—they are antianabolic and increase urea nitrogen to twice the baseline level.
- *Phenacetin*—it is nephrotoxic and puts added strain on an already damaged kidney.
- *Benzyl penicillin*—this has got significant potassium content and can be neurotoxic. So this drug is contraindicated in renal failure.
- *Others*—other drug which should be contraindicated are absorbed antacids, carbenicillin in large doses, ascorbic acid, ammonium chloride and laxatives.

Renal Osteodystrophy

It occurs due to defect in hydroxylation of 25-HCC to 1, 25-DHCC a process that normally occurs in kidney. Hypocalcemia occurs due to impaired calcium absorption and hyperphosphatemia, due to reduction in renal phosphorous excretion. Hypocalcemia results in secondary hyperparathyroidism with increased level of serum parathyroid hormone. Systemic acidosis is also associated with these conditions. There are symptoms of chronic renal failure.

Clinical Features

- *Myopathy*—muscle cramps are more common; ‘restless leg syndrome’, where patient’s legs jump at night.
- *Neuropathy*—sensory neuropathy may cause paresthesia. Motor neuropathy may present as foot drop. Autonomic neuropathy may cause delayed gastric emptying, diarrhea and postural hypotension.
- *Endocrine function*—there may be hyperparathyroidism and hyperparathyroidism. Amenorrhea is common in females. There is also loss of libido and sexual functions. Growth retardation occurs in children and bone fractures occur frequently.
- *CVS effects*—hypertension results in 80% of patients. This is due to sodium retention and increased secretion of renin, angiotensin-I and aldosterone. Atherosclerosis is common.
- *Acidosis*—there is also acidosis; cellular and humoral immunity are impaired.
- *Bone*—in adults, gradual softening and bowing of bone occurs.

Radiographic Features

- *Radiodensity*—generalized loss of bone density and thinning of bony cortex.
- *Lamina dura*—loss of lamina dura.
- *Angle of mandible*—thickness of cortex of mandibular angle is reduced.
- *Medullary spaces*—increase in medullary space at the expense of trabeculae. Other bones may show areas of sclerosis.

Diagnosis

- *Clinical diagnosis*—myopathy, neuropathy can give clue to diagnosis.
- *Radiological diagnosis*—generalized loss of bone density with thinning of bony cortex will aid in diagnosis.

Management

- *Vitamin D*—plasma Ca^{++} and K^+ are kept as near as normal.
- *Vitamin D*—hypocalcemia is corrected by giving hydroxylated synthetic analogues of vitamin D.
- *Phosphate binding agents*—hyperphosphatemia is controlled by dietary restriction of foods with high phosphate content (like milk, cheese, eggs) and the use of phosphate binding drugs.

Uremia

It is a clinical condition caused by the retention of urinary constituents in the blood. The characteristic symptoms are headache, itching, nausea, convulsions, and eventually coma. Patient’s breath may have urinous odor.

There is unpleasant taste and dryness of mouth. There is erythematous, pseudomembranous type of uremic stomatitis. If pseudomembrane is removed, it will expose dry, red, swollen mucosa. The oral bleeding tendency, often observed in patients with uremia, is due to uremic thrombopathy. Patient has an increased disposition to develop oral candidiasis.

Kidney Transplantation

In order to prolong life in patients with end stage renal failure, renal transplantation is done. It involves surgical removal of kidney from living first degree relative such as sibling, parent, child or a recently expired source. Kidney transplant patient usually receives a continuous regimen of immunosuppressive medication to ensure graft survival.

Clinical Features

- Majority of clinical manifestation occur secondary to immunosuppressive drugs.

- *Buffalo hump*—steroids are responsible for cushingoid effect, characterized by rapidly acquired adiposity about the upper portion of the body, mooning of face and tendency to become round shouldered and develop a 'buffalo hump' at the base of neck.
- *Infection*—there is increase susceptibility to fungal infection due to decreased migration and impaired phagocytic function of leukocytes and macrophages.
- *Hepatic dysfunction*—cyclosporine can cause hepatic dysfunction because metabolism occurs exclusively in the liver.

Oral Manifestations

- *Oral mucosa*—pale mucosa with diminished color demarcation between attached gingiva and alveolar mucosa.
 - *Salivary gland*—enlarged salivary glands and decreased salivary flow resulting in xerostomia.
 - *Odor and taste*—odor of urea on breath and metallic taste.
 - *Teeth*—enamel hypoplasia, dark brown stain on crowns, dental malocclusion, and low caries rate.
 - *Gingiva*—increase in calculus formation, low grade gingival inflammation and bleeding from gingiva and mouth.
 - *Candidiasis*—candidal infection can occur on tongue.
 - *Other*—petechiae, ecchymosis, erosive glossitis, burning and tenderness with dryness of mucosa. Dehiscence of wounds also occurs.
- *Defer dental treatment*—after transplantation, routine dental treatment should be postponed until maintenance dose; of immunosuppressive agents is reached.
 - *General guidelines for managing the kidney transplant patient who is taking corticosteroids:*
 - Always use long acting anesthetics agents such as bupivacaine.
 - Use mild sedatives for apprehensive patients. Use postoperative pain medications, when indicated.
 - Patients who are receiving alternate day corticosteroid therapy should be treated on off day.
 - If the patient is receiving daily therapy, then the dosage of oral steroids should be doubled the day before, the day of and 2 days after treatment.
 - Alternatively, single dose of 100 mg dose of hydrocortisone hemisuccinate should be given intramuscularly just before the treatment and extra 20 mg dose should be given early that evening.
 - *Following steps should be taken for proper management of renal transplant patients:*
 - The patient's physician should be consulted to coordinate treatment.
 - Broad spectrum antibiotics should be used in prophylactic medication of the patient, before dental treatment, because of decreased immune status.
 - Complete blood count with differential and platelet count should be obtained before any surgical procedure as corticosteroids can cause myelosuppression.

Radiological Features

- *Demineralization of bone*—there is demineralization of bone with loss of bony trabeculation.
- *Ground glass appearance*—sometimes, ground glass appearance can be seen.
- *Other finding*—loss of lamina dura, socket sclerosis is other radiological findings.

Diagnosis

Diagnosis can be made by history.

Dental Considerations

- *Elimination of source of infection*—Oral infection in transplant patients has been reported as frequently as pneumonia or urinary tract infection. Prompt diagnosis of the site and cause of infection in immunosuppressed patient should be on high priority. Identification of pathogens should be performed with culture, sensitivity, smear, aspiration technique or biopsy. The successful management of transplant patient begins before transplantation, with the preoperative elimination of potential sources of infection.

Disorders of Gastrointestinal Tract

Hepatitis

It is an acute, inflammatory and infective infection caused by hepatitis A, B, C, D and NANB viruses. Dentists are 3 to 4 times more likely to be exposed to hepatitis than with general population.

Types

- *Hepatitis A*—it is also called as *infectious hepatitis*. It is endemic and occurs in person who lives in poor living condition. Spread of hepatitis A is by orofecal route. The period of infectivity is highest during the week before the onset of clinical symptoms. It is common in children, primary and nursery school age. Once it occurs it gives life long immunity.
- *Hepatitis B*—it is also called *serum hepatitis*. It is transmitted by parenteral route, for example, blood and blood products and contaminated needles. It can also be transmitted due to close personal contact. It is also found in saliva, semen and vaginal secretions. 5 to 10%

of infected person remain as carriers. Hepatitis B tends to have greater mortality and morbidity than hepatitis A.

- **Hepatitis C**—it is caused by hepatitis C virus and it is responsible for sporadic viral hepatitis in intravenous drug user, and in patient with renal dialysis. It has got less clinical severity as compared to hepatitis B. It is associated with lichen planus, lymphoma, and cryoglobulinemia.
- **Hepatitis D**—it is called by delta agents or hepatitis D virus. It is incomplete virus carried within hepatitis B virus and will replicate in presence of HBsAg. So this infection occurs in association with hepatitis B infection. It is spread by parenteral route. It has got same clinical features as that of hepatitis B.
- **Hepatitis G**—it is caused by hepatitis G virus or GB virus-C. Many of this patient is infected with hepatitis C virus. It produces clinical symptoms less severe to hepatitis C.
- **Posttransfusion hepatitis**—this is caused by newer hepatitis virus like TTV (transfusion transmitted virus), SEN D and SEN H. These virus are not transmitted during dentistry.

Phases of Hepatitis

- **Prodromal phase**—is of 1-2 weeks; symptoms like anorexia, nausea, malaise and fever occur.
- **Icteric phase**—(6-8 weeks) anorexia, nausea, vomiting and pain in the right upper quadrant of abdomen. Hepatomegaly and splenomegaly may also be seen.
- **Convalescent (recovery) phase**—symptoms disappear, but abnormal liver function values may persist.

Clinical Features

- **Incubation period**—in this, most cases resolve completely within 4 months after onset of symptoms but, some end in fulminating disease and others progress to chronic hepatitis.
- **Symptoms**—systemic complains include malaise, arthralgia, morbilliform skin rash, anorexia, vomiting and myalgia. Patient has high grade fever with tenderness and enlargement of liver. Patient also complains of upper respiratory tract infection, distaste for cigarette, fever and enlargement of liver.
- **Signs**—jaundice, darkening of urine, splenomegaly and whitish stools occurs.
- **Oral manifestation**—icterus of the oral mucosa, which is seen on the palate and in the sublingual region.

Laboratory Findings

- **Plasma bilirubin level**—plasma bilirubin level excess of 3 mg/dl.
- **SGPT SGOT level**—SGPT, SGOT levels increase 10 times in hepatitis.

- **Alkaline phosphate level**—liver enzymes such as alkaline phosphatase and lactic dehydrogenase show slight elevation.
- **WBC**—WBC shows leukopenia, leukocytosis and atypical lymphocytes.
- **Australia antigen**—demonstration of HBsAg or Australia antigen.

Management

- **General treatment**:
 - **Symptomatic treatment**—like bed rest and prevention by isolation of blood, saliva contaminated objects; use of gloves and apron and sterilized instruments are must.
 - **Nutrition**—a high calorie diet should be given. It is usually given in morning because many patients experience nausea in evening.
 - **Drugs**—there is no specific drug useful for it. But interferon and ribavirin have been tried, with some success in chronic hepatitis.
 - **Prevention**—the patient should avoid salivary transmission to others by avoiding kissing, spitting and sharing food, cigarettes, utensils and sexual contact.
- **Hepatitis A**—it is self limiting and resolves with in one month mortality is very low. Treatment is usually symptomatic.
- **Hepatitis B**—chronic hepatitis B infection can be treated with lamivudine or interferon. Treatment is needed for 1 to 3 years.
- **Hepatitis C**—chronic hepatitis C is treated by combination of ribavirin, interferon alpha or pegylated interferon.
- **Hepatitis D**—drug treatment with alpha interferon is effective.
- **Hepatitis G**—alpha interferon is also effective against hepatitis G.

Prevention

- **Hepatitis A**—people who are known to have contact with a patient, such that they may have ingested minute amounts of fecal material or have been injected with as little as 0.0004 ml of infected blood, should be given prophylactic gamma globulin injections.
- **Hepatitis B**—a vaccine has been prepared from the plasma of asymptomatic carriers of hepatitis B. It is composed of non-infectious hepatitis B surface antigen particles. It is recommended in all high-risk groups.
- **Hepatitis C**—there is no vaccine available for this hepatitis. Preventing measure like not sharing personal item with bloodstain, not sharing needle in drug user, avoid having tattoo without strict health precaution.

- *Hepatitis D*—vaccination against hepatitis B will also give protection against hepatitis D.

Dental Considerations

- *Risk to dental professional*—hepatitis B, C and other types can be transmitted to the dentist by blood contaminated needles or instrument stick from an infected patient in acute phase of disease.
- *Clotting factors assessment*—if surgery is necessary, obtain preoperative prothrombin time and bleeding time, as in liver diseases deficiency of clotting factor may be present.
- *Universal infection precaution*—dental personnel may act as a source of infection to patient. Dentists who are carriers of HBV and who do not practice universal infection control precautions can transmit the infection to patient. At the same time it is required to avoid infection from patient to be transmitted to dental personnel.
- *Strict aseptic procedure*—strict aseptic procedure—use of masks, gloves for all persons is a must.
- *Minimize aerosol production*—minimize aerosol production by using a slow speed handpiece and using air syringe.

Inflammatory Bowel Disease

Chronic infectious diseases of small intestine and large bowel are of interest for dentist because of the oral manifestations that have been reported. In some of the cases it can be initial finding.

Types

- *Ulcerative colitis*—it is an inflammatory disease that is confined to the mucosa and submucosa of colon.
- *Crohn's disease*—it is an inflammatory disease involving the entire wall of a portion of small gut.

Clinical Features

- *Age*—it is seen in teenagers as compared to adults.
- *Symptoms*—upper and lower abdominal pain, usually cramping in nature along with fever and episodes of bloody diarrhea. Weight loss and malnutrition may occur in these patients.
- *Anorectal fistulae*—anorectal fistulae develop, as well as segmental narrowing of the intestinal lumen and rectal bleeding.
- *Erythema nodosum*—in some cases, there is presence of erythema nodosum of skin.

Oral Manifestations

- *Site*—the most frequently affected areas are the buccal mucosa, vestibule and lip.



Fig. 33-8: Linear ulceration in buccal vestibule with diffuse nodular swelling (Courtesy Dr Milind Chandurkar).

- *Aphthous ulcer like lesion*—Lesions resembling aphthous ulcer may occur.
- *Orofacial granulomatosis*—an oral granulomatous lesion occurs as nodular mass in mucobuccal fold.
- *Cobblestone appearance*—it shows cobblestone pattern of swelling on buccal mucosa.
- *Linear ulceration*—there is linear hyperplastic fold with ulceration (**Fig. 33-8**).
- *Swollen lip*—it appears swollen and indurated.

Diagnosis

- *Clinical diagnosis*—linear ulceration with granulomatous lesion with gastrointestinal symptoms may give clue to diagnosis.
- *Laboratory diagnosis*—biopsy shows non-necrotizing granulomatous inflammation.

Management

- *Sulfa drugs*—patient can respond to sulfasalazine. The metabolites of which are concentrated in the intestinal tissue.
- *Corticosteroids*—corticosteroids such as prednisolone can be use in combination with immunosuppressive drug azathioprine. Therapy is being used in increasing frequency in inflammatory bowel disease.
- *Management of oral lesion*—oral lesion are treated by topical and intralesional corticosteroids. Persistent lesions are treated with thalidomide.

Dental Considerations

- *Precaution*—it may necessitate alteration of dental therapy or special precautions on the part of dentist.

- *Corticosteroids therapy*—patients on corticosteroid therapy may develop both, hyperglycemia and osteoporosis, both of which have adverse effect on contemplated dental therapy.

Gastroesophageal Reflux Disease

Gastroesophageal reflux disease is a common condition. Regurgitation of gastric contents will reduce the pH of the oral cavity below 5.5. This acidic pH of oral cavity begins to dissolve enamel of teeth. It is most commonly seen on the palatal surfaces. Erosion of the enamel exposes the underlying dentin, which is a softer, opaque material. The extent of erosion depends on the frequency and the quantity of exposure along with the duration of disease. Newly exposed dentin is smooth and shiny, while dentin from previous exposures may be stained.

Erosion differs from dental caries in that it is a hard, dished-out area where enamel has dissolved and the underlying dentin is exposed. On the other hand, caries reveals soft, discolored dentin and results from the bacterial breakdown of sugars on the surface of the teeth.

The prevalence of caries is not increased in persons with GERD, possibly because the acidic environment interferes with the formation of the dental biofilm. Good dental care and control of acid helps decrease the prevalence of erosion. However, once the erosion occurs, it is irreversible and can only be treated with surgical restorative procedures. Therefore, early recognition and patient education is the most effective treat.

Jaundice

It is a symptom rather than a disease, resulting from excess of bilirubin in circulation. Jaundice can be recognized by color of skin, oral mucous membrane and sclera of the eyes. There is yellowish discoloration of skin, muscle membrane and sclera of eyes due to increased level of bilirubin and deposition of bile pigment tissues. Jaundice appears when serum concentration exceeds 2 to 3 mg /dl.

Types

- *Hemolytic jaundice*—results from an excessive hemolysis or destruction of erythrocytes produced by an inherited abnormality in the cells, some acute diseases and certain drugs or poisonous agent or acquired immune disease.
- *Obstructive jaundice*—caused by gallstone and external pressure on biliary passage, associated with infection or neoplastic lesions.
- *Hepatocellular jaundice*—it can be caused by diseases such as hepatitis and alcoholic and postnecrotic cirrhosis.

Causes

- *Hemolysis*—there is increased destruction of red blood cells with excessive pigment production.
- *Gilbert's syndrome*—it is a common form of congenital hyperbilirubinemia.
- *Transport disease*—inability of the liver to transport bilirubin into the bile.
- *Others*—jaundice can occur due to primary biliary cirrhosis, cystic fibrosis and traumatic biliary strictures.

Clinical Features

- *Urine*—increased urobilinogen excretion causes the urine to turn dark.
- *Anemia*—pallor occurs due to anemia and thus can be noticed on nails.
- *Splenomegaly*—it occurs due to excessive reticuloendothelial activity.
- *Others*—pruritus, abdominal pain and pale stools can also occur in jaundice.
- *Icterus*—yellowish discoloration of sclera of eye is also present. This yellow discoloration is caused by hypercarotenemia (**Fig. 33-9**).

Oral Manifestations

- *Icterus of oral mucosa*—it can be seen on palate and in the sublingual area.
- *Spontaneous bleeding*—in cases of severe jaundice patient may present with spontaneous bleeding in the oral cavity or severe bleeding following oral surgical or periodontal operation.

Diagnosis

- *Clinical diagnosis*—icterus and yellow discoloration of skin can easily be noted
- *Laboratory diagnosis*—serum bilirubin level is increased



Fig. 33.9: Icterus seen in eyes of patient who is suffering from jaundice. Note also yellowish discoloration of facial skin (Courtesy Dr Milind Chandurkar).

Management

- *Consult the physician*—if dentist discovers the jaundice patient should be sent to physician for further treatment and consultation.

Diseases of Esophagus

- *Plummer-Vinson syndrome*—It is characterized by an esophageal web with resulting dysphagia, particularly in the upper segment of esophagus, as well as by atrophic changes in the mucous membrane of mouth and by a hypochromic microcytic anemia.
- *Dysphagia*—it is difficulty in swallowing which may result from mechanical obstruction in the esophagus or from disorders of the nervous system that prevent the coordinated reflex contraction of the appropriate muscles.
- *Esophageal ulcer*—it can be associated with tetracycline therapy. It occurs more often after dry swallow of tablet or capsule, especially at bedtime. The patient complains of severe retrosternal burning pain.
- *Dental consideration*—due to unique position in treating in the oral cavity, dentist may be the first to detect this condition and should promptly refer it to physician for early diagnosis and prompt treatment. Special precautions should be taken while treating these diseases, to avoid aspirations secondary to dental procedures. Patients who have been prescribed tetracycline should be warned to swallow the capsule or tablet with adequate amount of liquid.

Peptic Ulceration

Ulceration of the mucosa of gastrointestinal tract caused by the action of protein-digesting pepsin on mucosa is one of the most common disease affecting GIT and one of the most common disease to afflict man.

Predisposing Factors

- *Acid production*—the normal stimuli for acid secretion include the thought, sight, smell or taste of food, which is mediated by the anterior hypothalamus, acts by vagal stimulation directly on the mucosa of stomach to cause acid production.
- *Hypoglycemia and mental stress*—hypoglycemia and mental stress also act by the same mechanism to increase stomach acid production.
- *Gastrin production*—presence of food in the antrum of stomach wall, by the mechanism of distention, cause increased production of the hormone gastrin, which acts directly on the acid producing cells of stomach.

Clinical Features

- *Site*—it usually affects lower one third of esophagus, stomach and the duodenum in increasing order of frequency.
- *Symptoms*—the most common symptom is epigastric pain, usually occurring either just before eating or 1-3 hours after eating. The pain is burning in nature and associated with nausea and vomiting. It is characteristically relieved by food. If ulceration is large enough to erode an artery, bleeding may be a primary symptom.
- *Coffee ground vomitus*—it is manifested by black tarry stools or more rarely by vomiting of blood. The vomitus appears like 'coffee ground because of blood reaction with acid.

Oral Manifestations

- *Site*—vascular formation is more commonly seen at the inner surface of the labial commissure and is more common in males than females.
- *Vascular malformation*—it is rarely manifested in the oral cavity but in certain cases, vascular formations of lip are also found in peptic ulcer. The vascular formations are of three types:
 - *Microcherry*—it is small, sharply circumscribed, red dot type of lesion.
 - *Glomeruli*—a conglomeration of tortuous, thin walled vessels 1-2 mm or more in size.
 - *Venous lakes*—dilated submucosal vein resembling a miniature varix.
- *Complication*—massive bleeding, obstruction, perforation and intractability to medical treatment.

Diagnosis

- *Clinical diagnosis*—coffee ground vomitus with vascular malformation of lip may give clue to diagnosis.

Management

- *Sedatives*—it is given to reduce mental stress, when it is the etiological factors.
- *Antacids*—it is given to neutralize the excess acid present in stomach. When antacids are given they usually are prescribed 1-3 hours after meals and at bedtime. Most antacids are combination of calcium carbonates, magnesium hydroxide and aluminum hydroxide.
- *Anticholinergic drugs*—it is given to decrease production of acids by the gastric mucosa.
- *H₂ histamine-receptor blocker*—such as cimetidine, famotidine, nizatidine or ranitidine. It blocks the action of histamine on the gastric parietal cells, thus reducing food stimulated acid secretion.

- *Sucralfate*—it act by covering and protecting the ulcer, thus promoting the healing.
- *Omeprazole*—it suppress gastric acid secretion by inhibiting an enzyme system at the secretory surface of the gastric parietal cells.

Dental Considerations

- *Drugs used*—the dentist should avoid administration of drugs that exacerbate ulceration, the most common of which is aspirin or one of its related compound. Patients who are given oral penicillin should be given penicillin V, instead of penicillin G, because of resistance of the former to gastric acid. Patient with anticholinergic drugs often present with dry mouth, which may present problems because of increase in the viscosity of mucus and discomfort in wearing complete dentures. There is also increased incidence of cervical caries. Many antacids contain calcium, magnesium and aluminum salts that bind both, erythromycin and tetracycline. These antacids may decrease the absorption of antibiotics by as much as 75 to 85%, if administered within 1 hour of such antacid therapy.
- *Sedation before treatment*—a patient who reacts in a particular stressful way to dental procedure should be sedated before dental treatment.

Neuromuscular Disorders

Auriculotemporal Syndrome

It is also called as *Frey's syndrome* or *gustatory sweating*. It is characterized by flushing and sweating of facial skin along the region of distribution of auriculotemporal nerve. It is an unusual phenomenon which arises as a result of damage to the auriculotemporal nerve.

Pathogenesis

- *Auriculotemporal nerve*—it gives sensory supply to the preauricular and temporal region, carries parasympathetic fibers to parotid gland.
- *Scar formation*—after damage to nerve scar formation occur around the nerve.
- *Re-established innervations*—the fibers regenerate, become misdirected and become connected with sympathetic nerve fibers of sweat gland and blood vessels of the facial skin.
- *Sweating and vasodilation*—parasympathetic fibers would therefore induce salivation; inadvertently stimulate the preauricular-dermal sweat gland and arterioles, causing vasodilation.

Variation of Frey's Syndrome

- *Chorda tympani syndrome*—it occur when there is injury to the submandibular gland.
- *Gustatory lacrimation syndrome*—this occur when there is injury to facial nerve proximal to geniculate ganglion.

Etiology

- *Surgical operation*—it follows surgical operations such as removal of a parotid tumor or ramus of mandible. It may follow superficial parotidectomy.
- *Parotitis*—parotitis of some type may damage the auriculotemporal nerve.
- *Congenital*—it may be due to birth trauma.
- *Drainage of abscess*—it may caused by inadvertent incision for drainage of parotid abscess.
- *Transaxonal excitation*—some cases of gustatory sweating appear due to transaxonal excitation, rather than actual anatomic misdirection of fibers.
- *Radical neck dissection*—some cases may develop after radical neck dissection.

Clinical Features

- *Age and sex*—there is no age and sex predilection.
- *Symptoms*—the patient exhibits preauricular flushing and sweating of the involved side of face, following ingestion of food or visual stimulation by food. Patient may sometimes feel pain while eating. The severity of sweating is increased by tart food. Profuse sweating may be evoked by parenteral administration of pilocarpine or eliminated by the administration of atropine. Local skin temperature can increase up to 2 °C.
- *Crocodile tears*—in it patient exhibits profuse lacrimation when food is eaten particularly hot and spicy food.
- *Cutaneous hyperesthesia*—presence of cutaneous hyperaesthesia in front and above the ear, the area supplied by the auriculotemporal nerve.

Diagnosis

- *Minor starch iodine test*—this test is done for the diagnosis. The test is as follows:
 - *Coating with 1% iodine*—preauricular area is coated with 1% iodine.
 - *Application of starch*—area is dried after application of iodine. After drying starch is applied on the skin.
 - *Sweating stimulant*—patient is given chocolate to induce sweating.
 - *Positive minor test*—bluish black discoloration is seen in area of sweating.

Management

- *Nerve dissection*—intracranial division of auriculo-temporal nerve has been reported to be successful.

- *Inserting physical barrier*—to prevent re-innervations of the sweat gland, many clinicians prefer to insert a physical barrier at the time of surgery. Materials used are porcine dermal collagen, autogenous tissue like fascia lata, temporalis muscle.
- *Pharmacological agents*—pharmacological agents used are atropine, scopolamine, and glucopyrrolate. It can be used topical or in injection form. Most commonly given injection is 1% glucopyrrolate lotion or cream.
- *Botulinum toxin*—this is the recent advance in the management of Frey's syndrome. BTX is the neurotoxin that blocks the release of acetylcholine by irreversible binding to presynaptic cholinergic autonomic nerve terminals.
- *Antimuscarinic agent*—systemic antimuscarinic agent oxybutynin chloride can be useful in treatment of Frey's syndrome.
- *Diluted formalin soaks*—the use of local skin application of diluted formalin soaks is well documented. It prevents hyperhydrosis. It act by blocking the effect of acetylcholine on sweat receptor, thereby inhibiting sweating locally. It should be applied before function to prevent embarrassing symptoms.

Bell's Palsy

It is also called as 7th nerve paralysis or *idiopathic facial paralysis*.

Pathogenesis

- *Normal course*—the cortical tract communicating with the motor nucleus ambiguous of facial nerve crosses over to get innervated into the lower face musculature. Upper face fibers are ipsilateral proximal to the nucleus.
- *Lower face palsy*—a cortical lesion will cause contralateral lower face palsy.
- *Total hemifacial palsy*—lesions of brainstem, main trunk or peripheral fibers will result in total hemifacial paralysis.

Etiology

- *Cold*—it usually occurs after exposure to cold. But many workers believe that it is a chance finding.
- *Trauma*—it may be a causative factor as Bell's palsy occurs after extraction of teeth and after injection of local anesthesia. Extraction and injection may cause damage to the nerve and subsequent paralysis.
- *Surgical procedure*—surgical procedures such as removal of parotid gland tumor in which the facial nerve is sectioned can also cause facial paralysis.
- *Ischemia*—it may caused by ischemia of the nerve near the stylomastoid foramen, resulting in edema of the

nerve, its compression in the bony canal and finally, paralysis.

- *Familial*—familial and hereditary occurrence is also reported in cases of Bell's palsy.
- *Facial canal and middle ear neoplasm*—these are usually associated with sensorineural hearing loss where 7th nerve palsy is a feature.
- *Tumors*—tumors of cranial base, parapharyngeal space and infratemporal fossa often cause 7th nerve palsy.
- *Other causes*—other causes like multiple sclerosis, atmospheric pressure change, and pregnancy can be triggering factors of facial paralysis.

Clinical Features

- *Age and sex*—women are more commonly affected than men and usually, it occurs in the middle age group. It arises more frequently in spring and fall, than at any other time of the year.
- *Onset*—it begins abruptly as paralysis of the facial musculature, usually unilaterally.
- *Prodromal symptoms*—in some cases, it is preceded by pain on the side of the face which is ultimately involved, particularly within the ear, temple, and mastoid area or at the angle of the jaw.
- *Symptoms*—speech and eating is difficult and occasionally, taste sensation on the anterior portion of tongue is lost or altered. Food is retained in the upper and lower buccal and labial folds due to weakness of buccinator.
- *Eye*—on the affected side, eye cannot be closed and wrinkles are absent on that side. There is watering of eye, which leads to infection.
- *Facial features*—when the patient smiles, the paralysis becomes obvious since the corner of the mouth does not rise nor does the skin of the forehead wrinkles or the eyebrows raise (**Fig. 33-10**).



Fig. 33-10: 7th nerve paralysis showing paralysis on left side of patient (Courtesy Dr Datarkar).

- *Mask like face*—the patient has a typical mask-like or expressionless appearance.
- *Drooling of saliva*—the muscular paralysis manifests itself by dropping of the corner of mouth, from which saliva may dribble.
- *Syndrome associated*—it is associated with Melkersson-Rosenthal syndrome.

Diagnosis

- *Clinical diagnosis*—slurred speech, mask like face, drooling of saliva can diagnose this condition.

Management

- *Vasodilator*—the use of vasodilator drug like histamine has been proved beneficial in some cases.
- *Surgical decompression and anastomosis of nerve*—surgical anastomosis of nerves has been carried out, especially with facial and hypoglossal nerve; thus can restore partial function.
- *Nicotinic acid*—administration of physiologic flushing dose of nicotinic acid.
- *Other*—systemic steroids or ACTH injection have been successful in treating Bell's palsy.

Motor System Disease

Motor system disease is neurodegenerative disorder which is characterized by progressive weakness and wasting of muscles.

Etiopathogenesis

- *Degeneration of motor neuron cells*—the motor system disease results from degeneration of the motor neuron cells of cranial nerves, anterior horn of spinal cord and pyramidal tract.
- *Genetic*—it is transmitted as autosomal recessive disorders.

Types

- *Progressive muscular atrophy*—it is seen in childhood.
- *Progressive bulbar palsy*—it is seen in children and young adults.
- *Amyotrophic lateral sclerosis (Lou Gehrig disease)*—it is seen in middle age.

Clinical Features

Progressive muscular atrophy

- *Age and sex*—it usually occurs in childhood. It shows a strong hereditary pattern, affects males more frequently than females.
- *Symptoms*—the initial symptoms usually consist of difficulty in walking with leg pain and paresthesia.

- *Steppage gait or stork-leg appearance*—patient notices atrophy of foot, leg and hand muscles ultimately occurs with the appearance of a typical foot-drop, steppage gait and stork-legs.

Progressive bulbar palsy

- *Age and sex*—it generally occurs in the children and young adults. There is no gender predilection.
- *Symptoms*—It is characterized by difficulty in swallowing and phonation, hoarseness, facial weakness and weakness of mastication. Chewing is difficult as facial muscles becomes weakened.
- *Signs*—atrophy of facial, masseter, temporal muscles and tongue, with fasciculation of the face and tongue. There is also impairment of palate and vocal cords.

Amyotrophic lateral sclerosis

- *Age and sex*—it generally occurs between the ages of 40 and 50 years and affects males more frequently.
- *Precipitating factors*—precipitating factors include fatigue, alcohol intoxication and trauma. Infections like syphilis, influenza, typhus and epidemic encephalitis can also lead to amyotrophic lateral sclerosis.
- *Symptoms*—the initial symptoms consist of weakness and spasticity of limbs, difficulty in swallowing and talking with indistinct speech and hoarseness.
- *Signs*—atrophy, flaccidity, symmetric weakness, slowness of movements, and impairment or loss of palatal movements may also occur.
- *Fasciculation*—there is small, synchronous, subcutaneous muscle contraction of the shoulders and thighs.
- *Tongue*—in earlier stages, the tongue is slightly weakened, leaving articulation relatively unaffected. In the middle stages, a gradual and generalized weakening of tongue occurs, accompanied by spasticity which results in reduced rate, range and force of articulatory tongue movements. In the later stages, there is virtually unintelligible articulation.

Diagnosis

- *Clinical diagnosis*—dysphagia, steppage gait, stork-leg appearance, tongue weakness and fasciculation are diagnostic point of motor neuron disease.

Management

- *Antiglutamate agents*—antiglutamate agent riluzole shows some improvement in this disease. As such there is no specific treatment for this disease and it is usually fatal.

Multiple Sclerosis

It is also called as *disseminated sclerosis*. This disease affects central nervous system. It is autoimmune disease.

Etiology

- *Allergic*—the lesions are allergic hypersensitivity manifestations of the nervous tissue due to antigen-antibody reactions.
- *Altered coagulation of blood*—the lesions are due to scattered venous thromboses in the nervous system associated with altered coagulation of blood.
- *Transitory local vasoconstriction*—the lesions are due to repeated, transitory localized vasoconstriction in various portions of the nervous system, precipitated by emotional disturbances or fatigue.

Clinical Features

- *Age and sex*—it occurs chiefly in younger age group with an onset of symptoms between the ages of 20 and 40 years. There is slight female predilection with familial occurrences.
- *Symptoms*—fatigability, weakness and stiffness of the extremities with ataxia or gait difficulty, involving one or both legs. Superficial or deep paresthesia. Personality and mood deviation towards friendliness and cheerfulness.
- *Signs*—variety of ocular disturbances including visual impairment as a manifestation of retrobulbar neuritis, nystagmus and diplopia.
- *Staccato speech*—staccato (a series of short, detached sound or words) type of speech.
- *Charcot's triad*—it consist of intentional tremors, nystagmus, dysarthria and scanning speech.

Oral Manifestations

- *Jaw weakness*—facial and jaw weakness occur in some patients.
- *Reduced mouth opening*—this may occur due to stiffness and weakness of muscle (**Fig. 33-11**).
- *Trigeminal neuralgia*—this occurs in multiple sclerosis patients and it usually bilateral.
- *Facial palsy*—this can also be present in the multiple sclerosis.



Fig. 33-11: Multiple sclerosis patient showing reduce mouth opening due to stiffness of facial muscle.

Diagnosis

- *Clinical diagnosis*—bilateral trigeminal neuralgia with speech difficulty and vision problems will diagnose this condition.

Management

- *Drug treatment*—beta interferon, mitoxantrone, and glatiramer can be effective in this patient. Medication to reduce muscle fatigue can also be used.
- *Physical and occupational therapy*—this can be use to patient up to certain extent.

Cerebral Palsy

The term cerebral palsy refers to a group of disorders with motor manifestations due to nonprogressive brain damage, occurring before or after birth.

Causes

- *Anoxia and ischemia*—anoxia and ischemia during labor can cause cerebral palsy.
- *Congenital infection*—congenital infection like toxoplasmosis, rubella, cytomegalovirus disease, herpes simplex, syphilis and influenza can also cause cerebral palsy.

Clinical Features

- *Forms*—cerebral palsy can be manifested in spastic, dyskinetic, ataxic or a combination, affecting one or four limbs.
- *Spastic form*—legs are commonly involved. Speaking problems with dysarthria, chewing and swallowing difficulty. Sometime, there are seizures associated with mental retardation.
- *Dyskinetic form*—it is characterized by athetotic purposeless movement, involving both agonist and antagonist muscles, which is increased by voluntary activity. Head movement and facial grimacing are characteristic.
- *Lead pipe type movement*—this occur due to excessive muscle tone.

Oral Manifestations

- *Sialorrhea*—drooling of saliva, this can lead to both functional and esthetic inconvenience.
- *Enamel hypoplasia*—an increased incidence of enamel defects.
- *Malocclusion*—this is present due to abnormal muscle behavior. Upper teeth are inclined.
- *Maxilla*—maxillary arch is tapered and ovoid in shape with high arch palate.

Diagnosis

- *Clinical diagnosis*—lead pipe type of movement with hemiplegia will lead to the diagnosis. There is also delay in development of motor skill.

Management

- *Physiotherapy* should be instituted as early as possible, in order to prevent contractures.
- *Orthopedic surgery* can be helpful in some cases.
- *Drugs therapy*—in case of seizures, *drugs therapy* like dantrolene sodium diazepam or L-dopa may be used.
- *Anti-cholinergic drugs*—for drooling of saliva, *anticholinergic drugs* such as benzhexol, atropine and scopolamine are used.

Epilepsy

It is a disorder, which results due to a sudden discharge by cerebral neurons, resulting in convulsive movements. It can cause sensory and motor abnormalities as well as loss of consciousness. Convulsions can also be seen with high grade fever, brain tumor, head injury, hypoglycemia, hypocalcemia, and drug toxicity.

Types

- *Grand mal*—it is most common type of seizure which can occur alone or with other types of seizure.
- *Petit mal*—it is the second most common type and it occurs without aura and with little or no clonic or tonic movements.
- *Psychomotor*—the seizures are preceded by an aura, which is often a hallucination or a feeling of déjà vu.
- *Jacksonian*—in it, attack begins in one part and spreads gradually.
- *Simple partial seizure*—it originates from one localized area of the brain and does not feature loss of consciousness.
- *Complex partial seizure*—it is same as psychomotor, with impairment of consciousness.
- *Status epilepticus*—it is the period of recurrent seizure attacks, without recovery between each attack.

Clinical Features

- *Grand mal*—it begins in childhood. There is warning followed by loss of consciousness.
 - *Aura phase*—a grand mal seizure begins with aura in which patient experiences epigastric discomfort, an emotion or hallucination of hearing, vision or smell.
 - *Tonic phase*—the aura is followed, in seconds to minute, by unconsciousness, cry, and tonic muscle spasms. There is also breathlessness due to spasm of respiratory muscles and the patient becomes cyanotic.

- *Clonic phase*—the tonic phase is followed by clonic phase composed of convulsive jerky movements, incontinence and tongue biting. Jaw is clamped shut and there is foaming at the mouth. Patient may injure himself, if he is near hard or sharp object.
- *Postictal state*—a postictal state is characterized by headache, confusion, lethargy, occasional temporary neurological deficit and deep sleep.
- *Petit mal*—they are present exclusively in children and frequently disappear during second decades of life. The patient loses his consciousness and appears to stare into the space. He will continue his normal activity immediately after the seizure.
- *Psychomotor*—during the seizure, the patients exhibit purposeless movements and bizarre behavior. Patient may wander about aimlessly, may get undressed or exhibit violent behavior during the seizure.
- *Jacksonian*—the seizure begins with clonic movement of a distal portion of extremities or the face. The convulsive movement spread up to the affected limb, becomes generalized and causing loss of consciousness.

Diagnosis

- *Clinical diagnosis*—tonic muscle spasm, convulsive jerky movement with headache and confusion will diagnose this condition.

Management

- *Immediate care of seizures*—you should move person away from danger like fire, water, machinery and furniture. After convulsion ceases it turns into recovery position, i.e. semiprone position. Do not insert anything in the mouth as tongue biting occurs at the start of seizure, so it cannot be prevented. If convulsion continues for more than 5 minutes or recur without person regains consciousness, summon urgent medical attention. Give intravenous anticonvulsants, i.e. diazepam, if convulsion continues or repeated. Airway should be kept patent during the epileptic fits.
- *Anticonvulsive drug therapy*—the drug of choice for grand mal, psychomotor and Jacksonian seizure is phenytoin (Dilantin), often in combination with phenobarbital. Patient resistant to it are given primidone. Drug of choice for petit mal is ethosuximide because of few side effects associated with it. Other drugs used are trimethadione, paramethadione and acetazolamide.

Dental Considerations

- *Gingival enlargement*—patients taking anticonvulsant drug such as phenytoin, develop gingival hyperplasia which may exacerbate due to local inflammatory factors. You should also look for level of drug in the gingival tissues and the effect of drug on gingival mast cells.

- *Side effects of phenytoin*—other side effects of phenytoin are megaloblastic anemia, lymphadenopathy, connective tissue and bone changes. Phenytoin blocks the effect of parathyroid hormone on bone, resulting in bone and root changes.
- *Drug interference*—aspirin, azole and metronidazole can interfere with phenytoin. So this drug is contraindicated in epilepsy.

Parkinson's Disease

It is a major cause of chronic disability in patients over 50 years of age.

Causes

- *Depletion of neurotransmitter*—it is caused by depletion of neurotransmitters, dopamine and norepinephrine in the basal ganglion.
- *Other disease*—some are caused by encephalitis, trauma, carbon monoxide intoxication, atherosclerosis, metal poisoning and brain tumor.

Clinical Features

- *Triad*—the triad is rigidity, tremor and bradykinesia (slowness in the initiation of movements).
- *Onset*—the onset is insidious. Mild stiffness of muscle of the extremities and tremor of hand are early signs.
- *Pill-rolling movement*—the typical hand tremors are called as pill-rolling movements and are caused by movement of thumb and fingers rubbing against one another.
- *Gait*—walking becomes more difficult and patients develop a slow shuffling gait in a stooped position because of the inability to stand straight.
- *Speech*—speech becomes slow owing to lack of muscle control (dysarthria) and as the disease progresses there is a decrease in all voluntary movements and increase in tremor.

Oral Manifestations

- *Appearance of face*—rigidity of facial muscle is common. The loss of flexibility gives the patient an expressionless or masklike face.
- *Droping*—the muscle rigidity also causes difficulty in swallowing, resulting in drooling.
- *Tremors*—tremor of tongue and mandible are also common, making speech and eating difficult for the patient and dental procedures difficult for the dentist.
- *Complication of Levodopa therapy*—due to Levodopa therapy, patient exhibits purposeless chewing, grinding and sucking movements that are quite bizarre. The patient may thrust or shake his tongue and chew or suck vigorously when there is nothing in mouth.

Diagnosis

- *Clinical diagnosis*—triad of tremor, rigidity and bradykinesia can be seen.

Management

- *Levodopa*—levodopa can cause dramatic reversal of symptoms of Parkinson's disease.
- *Anticholinergic drugs*—mild form of Parkinson's disease can be managed by anticholinergic drugs such as trihexyphenidyl, like benzotropine or ethopropazine.
- *Propranolol*—propranolol an adrenergic antagonist may be used to reduce tremors.
- *Bromocriptine*—bromocriptine is one of the newer drugs which are an agonist at the dopamine receptors.

Dental Considerations

- *Pretreatment sedation*—anxiety will increase both, tremor and degree of muscle rigidity. Pretreatment sedation with diazepam is often recommended, as it will reduce anxiety.
- *Precaution while changing posture*—when the dental treatment has finished, the patient should be warned to take care while changing from a supine to standing position, since levodopa has a significant orthostatic hypotensive effect.

Orofacial Dyskinesia

It is caused by extrapyramidal disorders, complication of phenothiazine therapy and dentures in gross malocclusion. It is more commonly occurs above 60 years.

It is characterized by severe involuntary, dystonic movements of the facial, oral and cervical musculature. Irregular and involuntary movements such as lip-smacking and lip-licking, protrusion of lips -as in pouting, protrusion of tongue and mandible with uncoordinated movements are mostly observed.

Correction of the denture occlusion may be effective therapy. Anti-Parkinsonism drugs therapy can be effective in some of the cases.

Granulomatous Disorders

Wegener's Granulomatosis

It is a disease of unknown etiology which basically involves the vascular, renal and respiratory systems. It is a granulomatous involvement of blood vessels resulting in necrosis of tissue. It is generally thought that the disease is aberrant hypersensitivity reaction to an unknown antigen.

Types (Clinical)

- *Generalized or classic Wegener's granulomatosis*—it involve upper respiratory tract, pulmonary and renal lesions.
- *Localized or limited Wegener's granulomatosis*—it affects oral and nasal cavity and the lungs.
- *Superficial Wegener's granulomatosis*—it exhibits lesion of skin and mucosa.

Clinical Features

- *Age and sex*—it usually occurs in 4th or 5th decade of life, with slight predilection for males.
- *Symptoms*—the most common symptom of Wegner's granulomatosis is nasal stuffiness with chronic discharge, which is sometimes bloody. Patient soon develops cough, hemoptysis, fever and joint pains. There is also presence of rhinitis, sinusitis and otitis or ocular symptoms. There are also non-specific symptoms of malaise, arthralgia and weight loss.
- *Signs*—hemorrhagic or vesicular skin lesions are also commonly present.
- *Renal symptoms*—glomerulonephritis, which develops ultimately to uremia and terminal renal failure.
- *Prognosis*—the disease is usually fatal, with mean survival time of 5 months. Death occurs due to involvement of kidney.

Oral Manifestations

- *Onset*—the disease usually starts with tumor like vegetations in mouth and nose (**Fig. 33-12**). Then inflammatory process starts in the interdental papilla, spreading rapidly in to the periodontium.
- *Ulceration*—ulceration can occur on any surface. Ulceration is usually perforating in nature.

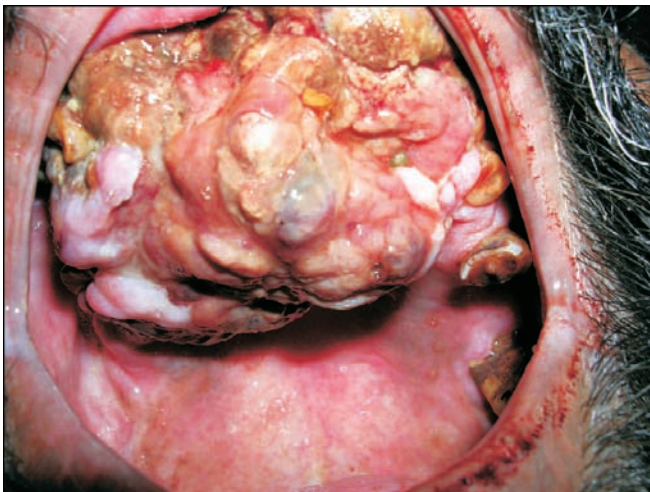


Fig. 33-12: Vegetative growth with ulceration in the palate in case of Wegener's granulomatosis (Courtesy Dr Parate).

- *Strawberry gingivitis*—involvement of gingiva is the most common manifestation; which is characterized by ulceration, friable granular lesions or simple enlargement of gingiva. Inflamed, hyperplastic appearing and hemorrhagic gingiva may be found. *Strawberry gingivitis* is the term used for this type of gingiva.
- *Palate*—oral lesions typically include ulceration of the palate by extension of nose lesions and destruction of nasal septum. This will lead to perforation of palate.
- *Teeth*—there may be loosening of teeth with in some cases spontaneous exfoliation. After extraction of teeth patient is usually noticed poor healing.
- *Other features*—cranial nerve palsies, jaw claudication, labial mucosal nodule, oroantral fistulae, and parotid swelling.

Radiological Features

- *Alveolar bone*—there are often signs of alveolar bone loss.

Diagnosis

- *Clinical diagnosis*—typical strawberry gingivitis with necrotic ulceration in the oral cavity.
- *Laboratory diagnosis*—cytoplasmic localization is present with Wegener's granulomatosis. Histopathologically chronic inflammatory cells and multinucleated giant cells are found.

Differential Diagnosis

Agranulocytosis, leukemia, lymphoma—diagnosis by blood picture, possibly histology.

Management

- *Cotrimoxazole*—it is combination of *trimethoprim* and *sulfamethoxazole*. It has proved to be effective as an adjuvant or sole therapy in both localized and generalized forms.
- *Corticosteroids*—regimen of cyclophosphamide 12 mg/kg body weight/day with prednisolone 1 mg/kg body weight have been utilized to obtain complete remission.
- *Others*—other treatment modalities includes cyclosporine, intravenous pooled immunoglobulin, and local irradiation.

Sarcoidosis

It is also called as *Boeck's sarcoid*, *Besnier-Boeck-Schaumann disease*. It is a disease of unknown etiology. It is a multi-system granulomatous disease. It is characterized by depression of delayed type of hypersensitivity, suggesting an impaired cell-mediated immunity and raised or abnormal serum immunoglobulin, suggesting lymphoproliferation.

Clinical Features

- *Age, sex and race distribution*—it commonly affects young adults with female predilection. It shows more prevalence in blacks.
- *Site*—lesions are most common in lungs, skin, lymph nodes, salivary glands, spleen and bones.
- *Symptoms*—mild malaise, fever, weight loss, fatigue and cough can be the chief features of the disease.
- *Signs*—there is presence of bilateral hilar lymphadenopathy, and pulmonary infiltration.
- *Lupus pernio*—these are the cutaneous lesions which appear as multiple, raised red violaceous patches. It occurs in group, grows slowly and does not ulcerate or crust.
- *Erythema nodosum*—it is nonspecific tender erythematous nodule seen on lower leg.
- *Eye lesion*—it includes anterior uveitis. In case of involvement of lacrimal gland patient may suffer from keratoconjunctivitis sicca.
- *Lofgren's syndrome*—it consist of erythema nodosum, bilateral hilar lymphadenopathy and arthralgia.

Oral Manifestations

- *Site*—it is rare in oral cavity, but cases are reported on salivary gland, lip, palate and buccal mucosa.
- *Salivary gland involvement*—it will lead to enlargement of salivary gland. This enlargement will lead to xerostomia patient. Due to xerostomia there is increased incidence of dental caries and ulceration of buccal mucosa.
- *Lip*—it appears as small papular nodules or plaques or resembles herpetic lesions or fever blisters.
- *Palate and buccal mucosa*—on the palate and buccal mucosa, it is described as bleb like, containing a clear yellowish fluid or as solid nodules.
- *Heerfordt's syndrome*—it consists of parotid enlargement, anterior uveitis, facial paralysis and fever.

Radiological Features

- *Jaw bone*—there is ill-defined radiolucencies which eroded the cortex. Expansion is absent in this case.
- *Chest radiograph*—radiological changes in lungs are of paramount importance. Based on chest radiograph, four stages of sarcoidosis can be distinguish:
 - *Stage 0*—characterized by a normal radiograph of the thorax.
 - *Stage 1*—bilateral enlargement of the hilar nodes, without pathologic changes in the lung fields. It represents an early stage of the disease.
 - *Stage 2*—bilateral enlargement of the hilar lymph nodes with pathologic changes in the lung fields around the hili.

- *Stage 3*—no enlargement of the lymph nodes, but extended and sometimes patchy or striped changes in both lung fields. The patches may be confluent and bullae may be formed.

Diagnosis

- *Clinical diagnosis*—salivary gland involvement with lymph nodes involvement with non-specific ulcer in the oral cavity, aids in clinical diagnosis.
- *Kveim-Siltzbach test*—is an intracutaneous test for the diagnosis of sarcoidosis—Kveim-Siltzbach test is positive in some cases. In this test, intradermal injection of a saline suspension of known human sarcoid tissue used as an antigen is given to a patient suspected to have sarcoidosis. One month after the injection, any palpable nodule is excised and examined histologically for evidence of a sarcoid reaction, or epithelial tubercle.
- *Laboratory diagnosis*—there is increased serum angiotensin converting level. Histopathologically *asteroid bodies* and *Schumann's bodies* are found.

Management

- *Corticosteroid*—asymptomatic patient requires no treatment as the lesion may resolve in 2 years of duration. In patient with symptoms corticosteroids can be given.
- *Refractory (does not respond to treatment) sarcoidosis*—in this case methotrexate, azathioprine, chlorambucil and cyclophosphamide is used.

Midline Lethal Granuloma

It is also called as *malignant granuloma*, *midline lethal granulomatous ulceration* and *midline nonhealing granuloma*. It is described as idiopathic progressive destruction of nose, palate, face and pharynx. Midline lethal granuloma is also associated with T cell lympho-proliferative disorders.

Etiology

- *Immune mechanism*—Lethal granuloma is due to a dysfunction of the immune mechanism normally responsible for granuloma formation.
- *Hypersensitive response*—Nowadays, it represents a fulminant hypersensitivity response to an unidentified antigen.
- *Vascular allergy*—vascular allergy like arthus phenomenon or periarteritis nodosa.

Clinical Features

- *Age*—it is more commonly seen in adults.
- *Site*—commonly involved sites are nose, palate, face and pharynx.

- **Onset**—it begins as superficial ulceration of the palate or nasal septum, often preceded by a feeling of stuffiness in the nose. This may persist for a month or two to several years. Pain may be present.
- **Progress**—eventually ulceration spreads from the palate to the inside of the nose and then to the outside.
- **Signs**—the palatal, nasal and malar bones may become involved, undergo necrosis and eventually sequester. Destruction is a prominent feature and loss of entire palate is common (**Fig. 33-13**). The patient may exhibit purulent discharge from the eyes and nose; perforating sinus tracts may develop and soft tissues of face may slough away leaving a direct opening into the nasopharynx and oral cavity.
- **Prognosis**—the patient ultimately dies of exhaustion or of hemorrhage if a large blood vessel is eroded. Death can also occur due to localized destruction of tissue in the midline of face.



Fig. 33-13: Destruction of palate seen in midline (Courtesy Dr Chole).

Diagnosis

- **Clinical diagnose**—destructive lesion in the midline of palate will diagnose this condition.
- **Laboratory diagnosis**—biopsy shows extensive necrosis with infiltration of some inflammatory cells and formation of occasional new capillaries.

Differential Diagnosis

- **Wegener's granulomatous**—in it there is presence of lung and renal diseases.
- **Noma**—in midline, lethal granuloma location is typical.

Management

- **Corticosteroids**—corticosteroid therapy has proven beneficial in some cases.
- **Radiation therapy**—radiation therapy of 5000 rad appears to be a treatment modality, in which remission of over 15 years is reported.

Chronic Granulomatous Disease

It is an uncommon hereditary disease with X-linked mode of transmission. It is characterized by severe recurrent infections as a result of defect of intracellular leukocytes enzymatic function, with decreased oxidative metabolism, in which there is failure to destroy certain catalase-positive microorganisms, including staphylococci.

Clinical Features

- **Age and sex**—majority of the patient are males, but females are also commonly affected. It is commonly found in children and infants.
- **Site**—there is widespread infection from infancy; usually affecting lymph nodes, lung, liver, spleen, bone and skin.
- **Skin lesion**—skin lesions occur on face, leading to tissue necrosis and granuloma formation.
- **Signs**—abscess, septicemia, pneumonia, pericarditis, meningitis and osteomyelitis are common features.

Oral Manifestations

- **Appearance**—granulomatous lesions of the oral and oropharyngeal mucosa usually present as sessile, lobulated, moderately firm and relatively nontender nodules and papules with normal coloration and with little or no surrounding inflammatory mucosal erythema.
- **Ulcerative lesion**—with time, some of the granulomas may ulcerate centrally and present as a deep, painless ulcer with a nonerythematous rolled border, reminiscent of squamous cell carcinoma.
- **Other features**—there is diffuse stomatitis, and benign migratory glossitis is also present.

Diagnosis

- **Clinical diagnosis**—granulomatous sessile lesion will give clue to diagnosis.
- **Laboratory diagnosis**—it is established by neutrophils function test.

Management

- **Conservative surgical excision**—localized lesions without systemic connection can be treated by conservative surgical removal and plastic surgical reconstruction.

- **Medication**—intralesional and systemic corticosteroids, low-dose radiotherapy, methotrexate, dapsone, salazosulfapyridine (sulfasalazine), hydroxychloroquine sulfate, among others. No therapy has proven to be universally effective in orofacial granulomatosis without systemic involvement.

Collagen Disorders

Scleroderma

It is also called as *systemic sclerosis*, or *Hidebound disease*. It is rare collagen disorder, which is characterized by hardening and tightening of the skin that can manifest as either localized or systemic form. It is a disease which involves connective tissue, blood vessels and lead to fibrosis. It is also called as progressive systemic sclerosis. Name is derived from sclero—hard derma—skin.

Types

- **Systemic or progressive systemic sclerosis**—it is diffuse and involves both skin and internal organ.
- **Localized form**—it involve the underlying muscle and bone along with skin and subcutaneous tissue. It is of two types:
 - **Morphea**—it is circumscribed form and is characterized by local changes limited to skin
 - **Linear**—it affects face, scalp and frontoparietal region.

Etiology

- **An endocrine dysfunction**—in some cases of endocrine dysfunction mainly thyroid and parathyroid disturbances can cause scleroderma.
- **Vascular disease**—basically an endarteritis obliterans, resulting in decreased vascular supply.
- **Nervous disorder**—since skin lesion often follows the distribution of nerves, nervous disorder may be causative factor.
- **Toxic or infectious agents**—shock or pneumonia, influenza, diphtheria and exanthematous disease.
- **Allergic reaction**—an antigen-antibody type of reaction is observed in scleroderma.
- **Environmental factors**—such as exposure to silica dust, vinyl chloride, benzene and tryptophan.

Clinical Features

Progressive systemic sclerosis

- **Age and sex**—it generally begins in childhood or young adult and greatest incidence is between 30 and 50 years of age. Females are more commonly affected with a ratio of 3:1.
- **Site**—it usually begins on face, hand or trunk.

- **Symptoms**—there is development of indurated edema of skin, neuralgia and paresthesia.
- **Signs**—Initial sign of PSS is frequently Raynaud's phenomenon, a paroxysm vasospasm of finger. In several months the edema is replaced by tightening and hardening of the skin, which results in difficulty in movements of the affected parts.
- **Involvement of internal organs**—hyperpigmentation, telangiectases and subcutaneous calcification may occur, leading to deformity and severe cosmetic problems along with involvement of internal organs.
- **Acro-steolysis**—resorption of terminal phalanges is also occur in scleroderma. Finger may also become clawlike.
- **Skin involvement**—skin area has thickened, hidebound cavity with lack of mobility of skin, limited mouth opening and renal involvement.
- **Syndrome associated**—it can be associated with progressive hemifacial atrophy (Parry-Romberg syndrome).

Localized form (circumscribed or morphea)

- **Site**—it usually occurs on the sides of the chest and thighs. They may be present for several months to many years.
- **Onset and progress**—it begins with violaceous patches on the skin. These lesions enlarge; become indurated and eventually loose hair and ability to sweat.
- **Hidebound character**—due to thickening patient assumes hidebound (constricted) character.
- **Signs**—progressively, these lesions turn into hypo or hyperpigmented areas depressed below the level of the skin. They may become stiff and hard. Affected skin appears rigid, inelastic, atrophic and shiny.

Localized form (linear)

- **Appearance**—a linear form of disease develops as a thin band of sclerosis that may run the entire length of extremities involving underlying muscle, bones and joints.
- **En coup de sabre**—a band made up of furrow with an elevated ridge on one side is often termed as 'coup de sabre' since it resembles the mark produced by the blow of saber (curved sword).

Oral Manifestations

- **Site**—the tongue, soft palate, lips and larynx are commonly involved.
- **Appearance**—these are characterized by mild edema, which is followed by atrophy and induration of mucosal and muscular tissue.
- **Mask like facies**—involvement of facial skin results in characteristic smooth, taut and masklike facies.
- **Mouse facies**—nasal alae may become atrophied resulting in pinched appearance to the nose resulting in mouse species.

- *Lips*—the lips become thin, rigid and partially fixed, producing microstomia.
- *Microstomia*—the oral aperture narrows considerably. Skin folds are lost around the mouth.
- *Tobacco pouch mouth or purse string appearance*—it can be seen periorally where furrow rows radiate from the atrophic vermilion borders, creating the so-called *tobacco pouch mouth or purse string appearance*.
- *Tongue*—tongue can become hard and rigid, losing its mobility and papillary pattern, making speaking and swallowing difficult. The color of tongue changes to a livid appearance. In the end stages, the tongue lays as a stiff, reduced body in the floor of mouth.
- *Lingual frenum*—the lingual frenum, which usually reflects the first oral change, shortens, becomes tendinous and finally disappears.
- *Neuropathy*—resorption of mandible may lead to pressure of the inferior alveolar nerve. This will result in neuropathy.
- *Dysphasia*—involvement of esophagus causes dysphagia.
- *Temporomandibular joint*—involvement of soft tissues around the TMJ leads to restricted movement of mandible, causing a pseudoankylosis.
- *Mandible*—when the facial tissues and muscles of mastication are involved the pressure exerted will cause resorption of mandible at the attachment of masseter muscle.
- *Salivary hypofunction*—in some cases salivary hypofunction can also be present.
- *Gingiva*—gingival hyperplasia may result from calcium channel blocker.

Radiographic Features

- *Periodontal ligament*—extreme widening of the periodontal ligament, two to four times the normal thickness, which is more prominent around posterior teeth with intact lamina dura. The periodontal changes are the direct result of changes within the membrane due to the underlying disease.
- *Margins*—borders of the lesions are smooth and sharply defined.
- *Bone resorption*—bone resorption occurs usually at the angle of mandible. It is bilateral and symmetrical.
- *Condylar involvement*—partial or complete resorption of condyle and coronoid process of the mandible, as a result of abnormal muscle pull or pressure from soft tissues is reported.

Diagnosis

- *Clinical diagnosis*—claw like finger, ulceration in finger, Raynaud's phenomenon, shiny skin, tobacco pouch mouth are typical features of this disease.

- *Radiological features*—periodontal ligament widening with resorption of bone at angle of mandible.
- *Laboratory diagnosis*—anticentromere antibodies or anti-Scl 70 are detected.

Management

- *D-penicillamine*—a drug has shown promise in the management by decreasing both, the skin thickening and organ involvement by interference with cross-linking of collagen and immunosuppression.
- *Extracorporeal photochemotherapy*—it is beneficial in skin lesion.
- *Other therapy*—other therapy like angiotensin converting enzyme inhibitor and calcium channel blocking agents may control hypertension in the patient.
- *Dental management*—the mouth opening may be increased by use of stretching exercises. Effective treatment of it is to use an increasing number of tongue blades between posterior teeth. If this is insufficient, bilateral removal of commissure may be necessary. Patient with extensive resorption of the angle of mandible is at risk of fracture and should therefore avoid trauma.

Kawasaki Disease

Kawasaki disease, or mucocutaneous lymph node syndrome, is a vasculitis that affects medium and large arteries with a corresponding cutaneous lymph node syndrome.

Clinical Features

- *Age*—it most commonly occurs in children between 3 months and 12 years of age.
- *Symptoms*—patients present acutely with edema, erythema of the hands and feet, fever and rash. The associated temperature must exceed 38.5°C (101.3°F) for 5 days to meet diagnostic criteria.
- *Signs*—bilateral congestion of ocular conjunctiva, indurative edema, erythema of palms, soles and membranous desquamation of fingers and toes. There is also polymorphous exanthema of torso without vesicles or crusts.
- *Lymphadenopathy*—acute nonpurulent swelling of cervical lymph nodes.
- *Cardiac sequelae*—cardiac sequelae to the vasculitis may result in aneurysm and myocardial infarction. Myocarditis commonly occurs within a week after the fever. Within 2-3 weeks, the previously edematous palms and soles peel and slough.

Oral Manifestations

- *Strawberry tongue*—strawberry like reddening and swelling of tongue papillae and diffuse reddening of

oral and pharyngeal mucosa, sometimes with gingival ulceration.

- *Lip*—the labia are cracked, cherry red, swollen, and hemorrhagic. The last of these may be due to the long-standing high-grade fever.
- *Facial palsy*—it is also some time seen in Kawasaki disease.

Diagnosis

- *Clinical diagnosis*—for diagnosis, 4 of the 5 following criteria must also be met: (1) peripheral extremity edema, erythema, or desquamation; (2) polymorphous exanthem; (3) bilateral conjunctival injection; (4) erythema and strawberry tongue in the oral cavity; and (5) acute cervical adenopathy.
- *Laboratory diagnosis*—there is proteinuria, leukocytosis, increased sedimentation rate and positive C-reactive protein.

Management

- *Intravenous gammaglobulin*—it is the most effective treatment in case of Kawasaki's disease.
- *Other drugs*—aspirin or systemic steroids can also be given in some cases.

Skin Disorders

Pityriasis Rosea

It is an acute skin eruption of unknown etiology.

Clinical Features

- *Age and sex*—it is more common in spring and autumn and it involves young adults chiefly, with no sex predilection.
- *Symptoms*—the lesion often manifests mild headache and low grade fever and cervical lymphadenopathy.
- *Primary lesions*—the generalized outbreak is frequently preceded by the appearance of a 'primary lesion' or 'herald spot' seven to ten days previously.
- *Appearance*—it is characterized by the appearance of superficial, light red macules or papules, generalized over most of the skin surface (**Fig. 33-14**).
- *Size*—the spot is bright-red and larger (3 to 4 cm in diameter) than the multiple eruptions which follow its appearance.
- *Shape*—the individual exanthematous lesion is commonly ovoid, with long axis parallel to the natural lines of cleavage of skin and are covered by a thin silvery scales.



Fig. 33-14: Superficial red macule seen over the skin in case of Pityriasis rosea (Courtesy Dr Pincha).

Oral Manifestations

- *Incidence*—the oral lesion occurs either concomitantly with, or subsequent to the skin manifestations.
- *Site*—it can occur on buccal mucosa, although tongue and palatal lesions have been reported.
- *Appearance*—oral lesion appears as erythematous macule, with or without central area of grayish desquamation.
- *Size and margins*—the lesion may be single or multiple, irregular in shape, occasionally showing raised borders and vary in size from few mm to 1-2 cm in diameter.

Diagnosis

- *Clinical diagnosis*—herald spot on the skin will give clue to diagnosis.
- *Laboratory diagnosis*—biopsy shows acanthosis and focal parakeratosis with microvesiculation or simply sprinkling of leukocytes within the epithelium.

Management

It requires no treatment since the disease is self limiting and generally undergoes rapid spontaneous regression.

Incontinentia Pigmenti

It is also called as *Bloch-Sulzberger syndrome*. It is transmitted as sex-linked dominant trait.

Clinical Features

- *Age and sex*—it appears shortly after birth and exclusively seen in females.
- *Appearance*—it is characterized by the appearance of erythematous and vesiculobullous lesions on the trunk and extremities which frequently disappear and

reappear. Then, they are gradually replaced by white keratotic, lichenoid, papillary or verrucous lesions, which then persist for some months.

- *Infant lesion*—some of the lesions in infants are brownish gray macules in a streaked, patchy distribution over the trunk and extremities, occurring subsequent to the verrucous keratotic lesions.
- *Melanin pigmentation*—there is heavy melanin pigmentation of epithelium, dropping down into cluster of chromatophores in the upper dermis (incontinence), which gives the disease its name.
- *Associated defect*—other defects can be seen like cataract, optic atrophy, strabismus, retrolental fibroplasia, central nervous system involvement and lesions of skeletal system.

Clinical Staging

- *Vesicular stage*—vesiculobullous lesion seen in trunk and limbs. Resolution within 4 months.
- *Verrucous stage*—verrucous cutaneous plaques develop. Resolution by 6 months.
- *Hyperpigmentation stage*—brown skin lesion with swirling (twisting) pattern.
- *Atrophy and depigmentation stage*—atrophy and depigmentation of skin can occur (Fig. 33-15).

Oral Manifestations

- *Site*—both the deciduous and permanent dentitions may be affected.
- *Teeth*—there is delayed tooth eruption, peg and cone shaped crowns, congenitally missing teeth, malformed teeth and additional cusps.



Fig. 33-15: Depigmented lesion seen on leg of patient in Incontinentia pigmenti (Courtesy Dr Pincha).

Diagnosis

- *Clinical diagnosis*—depigmented or vesicular lesion on trunk with cone shaped crown may suspect this disease.
- *Laboratory diagnosis*—biopsy shows intraepithelial clefts filled with eosinophils. It may show melanin containing macrophages.

Management

Dental management—dental defects can be corrected with the assistance of orthodontics and prosthetic dentistry.

Acanthosis Nigricans

It is acquired dermatological disorders characterized by velvety brownish discoloration of the skin.

Types

- *Benign*—it may be present at birth or occur later in childhood; appears to be genetic in origin inherited as a dominant characteristic.
- *Malignant*—it is associated with internal malignancy like adenocarcinoma of stomach and occurs in older age group.
- *Pseudoacanthosis nigricans*—it is most common and is associated with endocrinopathy.

Clinical Features

- *Site*—the most common areas involved are axilla, palms and soles and face and neck.
- *Appearance*—skin lesions are symmetric with mild hyperpigmentation and mild papillary hypertrophy of only small patchy areas. In some cases, it is heavily pigmented, aggressively verrucous lesion involving much of the skin.
- *Symptoms*—the verrucous lesions are often pigmented and generalized pruritis is also a common finding.

Oral Manifestations

- *Site*—the tongue and lips are most commonly involved, followed by buccal mucosa.
- *Incidence*—oral manifestations are most common with malignant form.
- *Tongue*—there is hypertrophy of the filiform papillae producing a shaggy, papillomatous surface on the dorsal tongue.
- *Lips*—the lips may be enlarged and covered by papillomatous growths, particularly at the angle of mouth.
- *Buccal mucosa*—the buccal mucosa may show a velvety white appearance with occasional papillary lesion.

- *Gingival hyperplasia*—gingival hyperplasia may also occur. Interdental gingiva may become so hyperplastic that it interferes with eating as it covers the teeth.

Diagnosis

- *Clinical diagnosis*—brownish alteration of skin velvety white appearance of buccal mucosa may be diagnostic features of scleroderma.
- *Laboratory diagnosis*—there is marked acanthosis, coupled with peculiar parakeratosis.

Management

Keratolytic lesion—this may improve appearance of benign form.

Ehlers-Danlos Syndrome

It is also called as *cutis hyperelastica* and it is a group of hereditary disorders of connective tissue. There is production of abnormal collagen, resulting in selective defect in collagen synthesis.

Clinical Features

- *Hyperelasticity*—there is hyperelasticity of skin, hyperextensibility of the joints and fragility of skin and blood vessels, resulting in excessive bruising as well as defective healing of skin wounds.
- *Rupture of large arteries*—rupture of large arteries as well as of intestine often occurs, producing a life threatening situation.
- *Rubber man*—in some patients, skin extensibility is pronounced, so that he can stretch the skin significantly. It is called as 'rubber man'.
- *Signs*—hypertelorism, a wide nasal bridge, epicanthic folds, protruding ears and frontal bossing are often present.
- *Subcutaneous nodules*—freely movable subcutaneous nodules are frequently found, which represent fibrosed lobules of fat.
- *Papyraceous scarring*—the scarring of skin following wound healing in these patients is unusual, as the scars tend to spread rather than contract in time. It resembles crumpled cigarette paper.

Oral Manifestations

- *Oral mucosa*—oral mucosa is of normal color but is excessively fragile and bruises easily.
- *Gingiva*—the gingival tissue appears fragile and bleeds after toothbrushing.
- *Gorlin sign*—it is positive in this patient. Patient can touch its nose with the tip of tongue.

- *Temporomandibular joint*—hypermobility of temporomandibular joint resulting in repeated dislocations of the jaw have been reported.
- *Teeth*—there may be lack of normal scalloping of the dentinoenamel junction, formation of irregular dentin and increased tendency to form pulp stones with hypoplastic changes in enamel.

Diagnosis

- *Clinical diagnosis*—hyperelasticity, hyperextensibility of joint and Gorlin's sign can give clue to diagnosis.
- *Laboratory diagnosis*—clotting time is normal, but capillary fragility test is usually positive.

Management

Not specific—there is no specific treatment for this disease, but surgical procedures should be carried out carefully as healing problems can exist.

Tuberous Sclerosis

It is also called as *Epiloia-Bourneville syndrome*. It is inherited as autosomal dominant trait.

Clinical Features

- *Facial angiofibroma*—it is also called as adenoma sebaceum. These are smooth surface papules (**Fig. 33-16**) which occur on nasolabial fold. Some lesions are also found under margins of nails.
- *Shagreen patches*—these are seen on skin and they resemble sharkskin-derived shagreen cloth.
- *Ash-leaf spots*—it appear on cutaneous surface.
- *CNS features*—it includes seizure, mental retardation. Potato like growth (tuber) can be seen at autopsy in CNS.
- *Cardiac rhabdomyoma*—it is tumor of heart. Myocardial function may hamper due to this tumor.



Fig. 33-16: Multiple papular facial lesions seen on face in case of tuberous sclerosis (Courtesy Dr Pincha).

- *Angiomyolipoma*—it can also occur in this syndrome and it is vascular smooth muscle adipose tissue tumor.

Oral Manifestations

- *Enamel defect*—there is developmental pitting defect on facial aspect of permanent dentition.
- *Fibrous papule*—it can also be present in this syndrome. These are most commonly occur in anterior gingival mucosa. In some cases diffuse gingival enlargement can also occur.

Diagnosis

- *Clinical diagnosis*—presence of facial angiofibroma, shagreen patch, mental retardation, and cardiac angiolipoma may diagnosis this disease. Gingival fibroma and enamel pit can also occur.
- *Laboratory diagnosis*—biopsy shows nonspecific fibrous hyperplasia in gingival fibroma.

Management

- *Anticonvulsant agents*—these are given to manage seizure.
- *Maintenance of oral hygiene*—as phenytoin is given in these cases, maintenance of oral hygiene is very important.

Seborrheic Keratosis

It is skin lesion which is characterized by benign proliferation of epidermal basal cells.

Clinical Features

- *Age*—it is common in older age group and seen in fourth decade of life.
- *Site*—it is seen on face, trunk, and extremities.
- *Appearance*—these are multiple, small tan to brown macule which enlarge gradually. They appear as stuck onto the skin.
- *Surface*—surface of lesion is fissured, pitted and verrucous.
- *Size*—size is less than 2 cm in diameter.
- *Dermatosis papulosa nigra*—it is seen in black people and is characterized by multiple, small dark brown papule scattered in zygomatic and periorbital region.
- *Leser-Trelat sign*—sudden appearance of seborrheic keratosis with pruritis has been associated with internal malignancy. This is called as Laser-Trelat sign.

Diagnosis

- *Clinical diagnosis*—small brown papular lesion seen on face and trunk will diagnose this condition.
- *Laboratory diagnosis*—there is exophytic proliferation of basilar epithelial cells with surface keratinization, acanthosis and papillomatosis.

Management

- *Cryotherapy*—cryotherapy with liquid nitrogen is used to remove the lesion.

Sebaceous Hyperplasia

It is localized proliferation of sebaceous gland of skin.

Clinical Features

- *Age*—it is common in older age group and seen in fourth decade of life.
- *Site*—it is seen most commonly on facial skin in the region of cheeks and forehead.
- *Symptoms*—it is usually painless.
- *Signs*—it is soft, nontender with yellow and white discoloration. There is small central depression which represents area where duct of sebaceous gland terminated.
- *Compression test*—after compression of lesion sebum (thick yellow product) is expressed from sebaceous gland.

Diagnosis

- *Clinical diagnosis*—compression test showing sebum from the lesion.
- *Laboratory diagnosis*—biopsy shows collection of enlarged sebaceous gland around the centrally located sebaceous ducts.

Management

- *Excisional biopsy*—this is done to differentiate it with basal cell carcinoma.

Actinic Lentigo

Lesion which occurs due to ultraviolet light damage to skin. It is also called as *lentigo solaris*, *solar lentigo*, *age spot*, *liver spot* and *senile lentigo*.

Clinical Features

- *Age*—it is common in older age group, in the age group of 40 to 70 years.
- *Site*—it is common on dorsal surface of hand, face and arms. Commonly seen in white people.
- *Appearance*—it is multiple with individual lesion appear as uniformly pigmented brown to tan macule with well demarcated and irregular border.
- *Size*—size is less than 5 mm.

Diagnosis

- *Clinical diagnosis*—brown to tan pigmented macule on face may give clue to diagnosis

- *Laboratory diagnosis*—elongated and club-shaped rete pegs are present.

Management

- *Topical retinoic acid*—this can reduce the color intensity of lesion.
- *Laser*—Q shaped ruby laser can be use for the treatment of this lesion.

Melasma

It is symmetric hyperpigmentation of sun exposed skin, face and neck. This is usually associated with pregnancy so it is also called as mask of pregnancy.

Clinical Features

- *Age*—it is seen in adult women.
- *Site*—it is seen in midface, forehead, upper lip, and chin.
- *Appearance*—it is bilateral, light to dark brown cutaneous macule. Pigmentation may darken with time.
- *Size*—size range from few millimeter to 2 cm in diameter.

Diagnosis

- *Clinical diagnosis*—bilateral symmetric pigmentation in pregnancy patient will diagnose this condition
- *Laboratory diagnosis*—there is increase melanin deposition in epidermis.

Management

- *Three percent hydroquinone and tretinoin*—it is effective in many cases.

Xeroderma Pigmentosum

It is genodermatoses in which multiple cutaneous malignancies develop. It is inherited as autosomal recessive trait and caused by defect in the excision repair mechanism of DNA.

Clinical Features

- *Age*—it occurs in early age group.
- *Symptoms*—there is increased tendency to sunburn. Patient also notices freckled pigmentation and patchy depigmentation of skin. Patient also notices neurological manifestation like below normal intelligence.
- *Actinic keratosis*—it is evident in early age group. These lesions will change into basal cell carcinoma or squamous cell carcinoma.
- *Oral features*—squamous cell carcinoma develops on lower lip and tongue.

Diagnosis

- *Clinical diagnosis*—skin cancer in young individual will give strong clue to diagnosis of Xeroderma pigmentosum.
- *Laboratory diagnosis*—biopsy shows features of cutaneous malignancies.

Management

- *Topical chemotherapeutic agents*—topical chemotherapeutic agent's 5-fluorouracil is used to treat actinic keratosis.
- *Genetic counseling*—it should be done before marriage.
- *Prevention*—patient is ask to avoid sun exposure, wear protective clothing

Muscle Disorders

Muscular Dystrophy

It is genetically determined disease characterized by degeneration of muscle leading to progressive weakness.

Types

- *Severe generalized familial muscular dystrophy*—it is described as a rapidly progressive muscle disease, usually beginning in early childhood and presenting a strong familial transmission.
- *Mild restricted muscular dystrophy*—it is a slowly progressive proximal myopathy which primarily involves the muscles of shoulder and face and has a weak familial incidence. It is transmitted as an autosomal dominant trait.

Clinical Features

- *Age and sex*—it predominately affects males. It begins in childhood, usually before the age of 6 years and rarely after 15 years.
- *Symptoms*—the earliest symptom is inability to walk or run due to which, the children fall readily and is associated with muscular enlargement and weakness. In case of mild restricted dystrophy, patient is unable to raise arms above the head and inability to close eyes.
- *Waddling gait*—the muscular enlargement ultimately proceeds to atrophy and the limbs appear flaccid. It is the atrophy which is responsible for the postural and ambulatory defects, such as *waddling gait*.
- *Signs*—scapular muscles become atrophic and weak with subsequent alteration in the posture.
- *Cardiac abnormalities*—cardiac abnormalities including cardiomegaly and tachycardia are often present and many patients die of sudden cardiac failure.

Oral Manifestations

- *Location*—the muscles of mastication, facial ocular, laryngeal and the pharyngeal muscles are usually involved, only in the late course of disease.
- *Teeth*—due to lack of muscle tension, teeth cannot be kept properly aligned in the arch.
- *Tapir-lips*—the lips develop a characteristic looseness and protrusion, which have been described as ‘tapir-lips’. The patient is unable to whistle or smile.
- *Open bite and diastema*—there may be severe open bite and development of diastema.
- *Temporomandibular joint*—locking and clicking of the jaw.

Diagnosis

- *Clinical diagnosis*—there is muscular weakness, waddling gait, tapir-lip will give clue to diagnosis.
- *Laboratory diagnosis*—there is gradual disappearance of muscle fibers, as the disease progresses no fibers may be recognized. Serum creatinine phosphokinase levels are elevated in all males.

Management

There is no treatment for this disease. Physical therapy may help prolong the use of specific muscle group.

Myotonias

Myotonias are the disorders characterized by abnormally slow relaxation after muscle contraction.

Types

- *Dystrophic myotonia*—it is also called as *myotonic dystrophy* or *dystrophic myotonia*. It is inherited as an autosomal dominant trait.
- *Myotonia congenita (Thomsen’s disease)*—it is transmitted as an autosomal dominant trait with incomplete penetrance in some families. It is generalized myotonia without weakness.
- *Myotonia congenita (Becker type)*—it appears late in childhood and characterized by muscle hypertrophy.

Clinical Features

- *Location*—atrophy of the muscles is seen usually in the hands and forearms. It can be seen in muscles of face, jaws, neck and levator of eyelids.
- *Symptoms*—there is associated weakness of the muscles. Muscular contraction induces a severe, painless muscular spasm, and an actually delay in relaxation.
- *Facial muscle*—alteration in the facial muscles, which consist of ptosis of the eyelids and atrophy of the masseter and sternocleidomastoid muscles.

- *Myopathic facies and swan neck*—the masseteric atrophy produces narrowing of the lower half of the face which, with ptosis and generalized weakness of the facial musculature gives the patient a characteristic ‘myopathic facies’ and ‘swan neck’.
- *Percussion contraction*—electrical and physical stimulation of a muscle produces the characteristic prolonged contraction or ‘percussion contraction’.
- *Dysphagia*—pharyngeal and laryngeal myotonia also exhibit weakness manifested by a weak, monotonous nasal type of voice and subsequent dysphagia.
- *TMJ*—recurrent dislocations of the jaw is also reported in this disease.
- *Eye*—blinking with strong closure of the eyes will sometimes produce a prolonged contraction of the lids.
- *Other*—other features are testicular atrophy, cataract, hypothyroidism with cold extremities, slow pulse, loss of hair and functional cardiac changes.

Diagnosis

- *Clinical diagnosis*—swan neck, muscular weakness, myopathic facies and atrophy of muscles of mastication.
- *Laboratory diagnosis*—there is enlargement of scattered muscle fibers and the presence of centrally placed muscle nuclei in long rows. True hypertrophy in some fibers, is found, as well as in isolated fibers which show extreme degenerative changes including nuclear proliferation, intense basophilic cytoplasmic staining and phagocytosis.

Management

There is no specific treatment for this disease.

Hemifacial Spasm

It is a disease characterized by a repeated, rapid, painless, irregular, nonrhythmic, uncontrollable, unilateral contraction of the facial muscles. It may occur due to compression of the facial nerve, in the facial canal, adjacent to the stylomastoid foramen.

Clinical Features

- *Location*—it usually begins in the periorbital muscles, but soon spreads to the entire half of the face.
- *Precipitating factors*—these spasms are often triggered by fatigue, tension or facial activity and are of brief duration, usually lasting for only a few seconds.
- *Symptoms*—it is first manifested as a brief transitory twitching, but may progress to sustain a spasm.
- *Signs*—in cases of long standing hemifacial spasms, mild facial contracture may occur with lid closure and lip pursing.

Diagnosis

- *Clinical diagnosis*—muscular spasms which is triggered by fatigue and twitching.

Management

- *Decompression of the nerve*—decompression of the nerve in its canal has offered relief in some cases. Prognosis is good, remission and recurrence may occur over a period of years.

Paramyotonia

It is a nonprogressive myotonia, inherited as an autosomal dominant characteristic that is not associated with muscular wasting. Characteristically, the cramping attacks are precipitated on exposure to cold.

Clinical Features

- *Symptoms*—it is manifested by cramping, stiffness and weakness of the muscles of the face and neck, fingers and hands, upon exposure to cold. Muscles cramping may disappear within an hour; the weakness may persist for several days.
- *Mask-like appearance*—the eyelids are closed and the face assumes a *mask-like* appearance.
- *Tongue*—the tongue may exhibit a similar cramping after drinking cold liquids and the speech becomes blurred. In many cases, myotonia of the tongue may be induced by percussion.

Diagnosis

- *Clinical diagnosis*—masklike appearance seen in face. Muscle cramping will disappear within hour after exposure to cold.

Management

There is no specific treatment for it, but the prognosis is excellent with frequent improvement during adult life.

Myasthenia Gravis

It is an autoimmune chronic disease characterized by progressive weakness of the skeletal muscles, particularly those innervated by the cranial nerves. It is characterized by easy fatigability of the striated muscles secondary to disorders at the neuromuscular junction. It affects acetylcholine receptors of muscle fibers resulting in fatigability of skeletal muscle.

Etiology

- *Defective neuromuscular transmission*—there is a defect in the neuromuscular transmission, which occurs due to

coating of acetylcholine receptors by circulating antibodies. This will result in the fault acetylcholine mechanism.

- *Hyperplasia of thymus*—it may occur in the endocrine system due to thymus hyperplasia or tumors of the thymus.
- *Other factors*—it may be related to pregnancy, menstruation and hyperthyroidism.

Clinical Features

- *Age and sex*—it occurs in adults in the middle age group with a predilection for women.
- *Symptoms*—there is rapidly developing weakness in voluntary muscles, following even minute activities. Patient may suffer from diplopia (double vision) and ptosis (drooping eyelids) and extraocular muscular paresis (an inability to focus the eyes) and dysarthria (slurring of word).
- *Sorrowful appearance of face*—there is dropping of the face, leads to sorrowful appearance of the patient.
- *Signs*—the neck muscles may be so weak that the head can not be held up without support.
- *Progress*—patients become exhausted, loses weight becomes further weakened and may eventually become bedridden. Death frequently occurs from respiratory failure.

Oral Manifestations

- *Symptoms*—the patient's chief complaints may be difficulty in mastication, deglutition and dropping of the jaw. Speech is often slow and slurred and disturbance in taste sensation occurs. Dysphagia and regurgitation of food are common.
- *Tongue*—there may be weakness of the tongue and palatal muscles. Protrusive movements of the tongue may become weak leading, at times to posterior collapse of the organ with airway obstruction.

Diagnosis

- *Clinical diagnosis*—ptosis, diplopia, dysphagia, sorrowful appearance of face.
- *Laboratory diagnosis*—elevated serum AChR (acetylcholine) level. Biopsy shows focal collection of small lymphocytes or *lymphorrhages* around small blood vessels in the interstitial tissue of the affected muscles.

Management

- *Anticholinesterases*—pyridostigmine, edrophonium, and neostigmine, administered intra-muscularly improve the strength of the affected muscles few minutes.
- *Corticosteroids*—these can be used in combination with cholinesterase inhibitors.

- *Thymectomy*—this is done in condition when myasthenia gravis is associated with thymoma.
- *Plasmapheresis* has been temporary valued in patient with severe exacerbation.

Dermatomyositis

It is also called as '*polymyositis*'. It is an acute or a chronic disease of unknown etiology and is characterized by gradual onset with vague and indefinite prodromata, followed by edema, dermatitis, myositis and sometimes neuritis and mucositis.

Clinical Features

- *Age and sex*—it may occur in patients of any age ranging from very young children to elderly, but majority occurs in the 5th decade of life. There is no sex predilection.
- *Onset*—it begins with erythematous skin eruptions, edema, tenderness, swelling and weakness of the proximal muscles of limbs. Fever may be associated with it.
- *Progress*—the weakness of muscle is progressive and characteristically spreads to face, neck, larynx, pharynx and heart.
- *Skin*—the skin becomes the seat of violaceous erythema and edema with a predilection for the eyelids, malar area and dorsa of hands. The typical skin lesions include *heliotrope* (liac-colored) changes around the face and fingers.
- *Signs*—the edema which gives the skin a puffy consistency including the face, leaves a reticulated telangiectatic erythema when it subsides.
- *Calcinosis cutis*—the skin lesions frequently calcify and form calcium carbonate nodules with a foreign body reaction which is known as calcinosis cutis.
- *Calcinosis universalis*—the term calcinosis universalis is applied when these calcified masses are found generalized throughout the soft tissues.
- *Prognosis*—muscle involvement may become severe enough to confine the patient to bed or cause death owing to failure of respiratory muscle.

Oral Manifestations

- *Diffuse stomatitis and pharyngitis*—the oral lesions consist of diffuse stomatitis and pharyngitis and are extremely common.
- *Symptoms*—involvement of the muscles of jaw, tongue and pharynx may pose problems in eating and phonation.
- *Oral mucosa*—the oral mucosa may show dark red or bluish erythema.
- *Tongue*—in the early stages, tongue is swollen and later becomes harder and gradually it becomes atrophic. The tongue may become rigid owing to severe calcinosis.

- *Lips*—telangiectatic lesions of vermillion border of lips and cheeks may also occur.
- *Teeth*—there is purplish black intrinsic staining of teeth.

Radiographic Features

- *Pulp calcification*—there is severe calcification and obliteration of pulp chambers of deciduous and permanent teeth.

Diagnosis

- *Clinical diagnosis*—heliotrope lesion with dark red oral mucosa, calcinosis cutis will give clue to the diagnosis.
- *Radiological diagnosis*—pulp calcification and obliteration is present.
- *Laboratory diagnosis*—the muscle fibers in dermatomyositis exhibit widespread degeneration and hyalinization. Many fibers show vacuolization, granulation and fragmentation with phagocytosis of disintegrating fibers. There is also mild anemia or leukocytosis. In addition, creatinuria is a constant finding as well as elevated levels of serum transaminase and aldolase.

Management

- *Corticosteroids*—corticosteroid should be given but its effectiveness is doubted.

Immunological Disorders

Primary Immune Deficiency

These are hereditary abnormalities characterized by an inborn defect of the immune system. These diseases may involve the B-cell system, T-cell system or a defect in both.

Types

- *Sex linked agammaglobulinemia*—it is caused by a defect in B-cell function with the T-cell function remaining intact. Due to this, these patients lack the ability to synthesize all classes of antibodies including the secretory immunoglobulin, making them more susceptible to bacterial infection. The symptoms begin at 6 months of age.
- *Primary adult immunoglobulin deficiency*—it is characterized by an abnormality of the B-cell or humoral antibody system that does not become clinically apparent until adulthood. Usually one or two Ig classes are deficient. The selective deficiency accounts for the relatively asymptomatic nature of the disease throughout childhood.
- *Thymic hypoplasia*—it consists of *DiGeorge's syndrome* and *Nezelof's syndrome*. These patients have normal

levels a serum immunoglobulin but lack cell mediated immunity. It consists of both, abnormalities of thymus and parathyroid glands. The lack of parathyroid hormone will lead to hypocalcemia and tetany; lack of thymus function causes absent T-lymphocyte response.

- *Secondary combined immunodeficiency*—it can be inherited as either sex-linked or an autosomal recessive trait. In these diseases, there is both T-cell as well as B-cell deficiency. The patients have low peripheral lymphocytes count severe deficiency of immunoglobulin and complete lack of cellular immunity.
- *Immunodeficiency with ataxia telangiectasia*—it is inherited as an autosomal recessive trait. There is combined T-cell and B-cell deficiency. Progressive neurologic disease form severe degenerative changes on the cerebellum, leading to cerebellar ataxia. It becomes apparent when the child begins to walk.

Clinical Features

- *Sex-linked agammaglobulinemia*—patients experience severe recurrent bacterial infections of lungs, meninges, skin and sinuses. There is hypoplasia of lymph nodes, adenoid and tonsils. There is increased incidence of rheumatoid arthritis, dermatomyositis, lymphoma and leukemia.
- *Primary adult's immunoglobulin deficiency*—the most common symptoms include recurrent gram-positive bacterial infections of upper and lower respiratory tracts.
- *Thymic hypoplasia*—there is increased susceptibility to infections with virus and fungi. Infection with *Candida albicans* is especially prominent.
- *Severe combine immunodeficiency*—symptoms begin in first few week of life and include bacterial, viral and fungal infections. Localized and systemic candidiasis is common. The patient dies of overwhelming infections during the first year of life, unless a histocompatible relative is found for a bone marrow transplant.
- *Immunodeficiency with ataxia telangiectasia*—telangiectasia of the skin and eyes becomes apparent at about 3 years of age. The lesion becomes more extensive with age and mainly occurs on conjunctiva, ears and malar eminences. These are also associated with Gonadal dysgenesis and increased incidence of malignancies of lymphoreticular system.

Oral Manifestations

- *Infection*—in T-cell deficiency, there may be chronic oral candidiasis and herpes simplex infection. In B-cell deficiency, there are recurrent bacterial infections and chronic maxillary sinusitis.

- *Congenital defects*—congenital defects like cleft palate, micrognathia, bifid uvula and short philtrum of upper lip.
- *Oral ulcerations*—occasionally, oral ulceration is seen in these patients but it is not the diagnostic of these disorders.

Diagnosis

- *Clinical diagnosis*—increase susceptibility to infection and severe recurrent infection will suspect immune deficiency.

Management

- *Control of local infection*—minimize the chances of local infection or septicemia and oral candidiasis, treated with antifungal therapy, prior to dental treatment.
- *Concentrated human gamma globulin*—patient with symptomatic B-cell abnormalities are usually given continuous therapy with concentrated human gamma globulin. When oral surgery is necessary, an extra dose of gamma globulin should be administered the day before surgery.
- *Blood replacement therapy*—patients with B cell deficiency resulting in the absence of particular immunoglobulin may experience severe transfusion reaction when receiving blood from a patient who has normal immunoglobulin levels. The immunoglobulin acts as a foreign protein and causes an allergic response. For these reasons, patient with selective IgA deficiency must be given IgA depleted blood in blood replacement therapy.

Miscellaneous Conditions

Drug Allergy

Allergy or hypersensitivity is an unwanted response of the body to the complete dose of drug.

Mechanism

- *Formation of antibody*—a patient previously exposed to a drug or other antigen has antibody, primarily IgE, fixed to basophils and mast cells.
- *Releasing of active mediators*—when the antigen in the form of a drug, food or airborne substance is reintroduced into the body it will react with the fixed antibody, bind complement and open the mast cells releasing active mediators such as histamine and slow reactive substance of anaphylaxis.
- *Vasodilation and increases capillary permeability*—these substances cause vasodilation and increased capillary

permeability resulting in fluid and leukocytes leaving the blood vessels and accumulating in the tissue forming areas of edema.

- *Constriction of bronchial smooth muscle*—constriction of bronchial smooth muscle also may result, when IgE is bound in the pulmonary region.

Clinical Features

Localized anaphylaxis

- *Urticaria*—when it involves the superficial blood vessels, urticaria (hives). It begins with pruritus in the area of release of histamine and other active substances.
- *Skin*—wheals (welts) then appear on the skin as an area of localized edema on an erythematous base.
- *Edema*—there is also macular papular or nodular rash with edema of skin and subcutaneous tissue.

Angioneurotic edema

- *Causes*—it can be caused by contact with an allergen or can be idiopathic. A recurrent form is inherited as an autosomal dominant trait. There is deficiency of an alpha-2 globulin, which normally acts as an inhibitor of the first component of complement and kallikrein.
- *Locations*—Angioneurotic edema of lips, tongue, eyelids, larynx and bronchi may occur.
- *Appearance*—in angioneurotic edema, when deeper blood vessels in the subcutaneous tissue are affected, a large diffuse area of subcutaneous swelling is produced under the normal overlying skin.
- *Respiration*—it is temporarily disfiguring, but not serious, unless the posterior portion of the tongue or larynx compromise respiration.

Serum sickness

- *Causes*—it frequently occurs after administration of foreign serum, which before antibiotics, was given for the treatment of infectious disease. It occurs from tetanus antitoxin, rabies antiserum and drugs that combine with body proteins to form allergens.
- *Mechanism*—in it, antibodies form immune complexes in blood vessels with administered antigens. The complexes fix complement, which attract the leukocytes to the area causing direct tissue injury.
- *Symptoms*—major symptoms consist of fever, swelling, lymphadenopathy, joint and muscle pain and rash.
- *Signs*—less common manifestation include peripheral neuritis, kidney disease and myocardial ischemia.

Generalized anaphylaxis

- *Mechanism*—it is an allergic emergency with no time to call consultants. It is reaction of IgE antibodies with an allergen, causing the release of histamine, bradykinin and SRS-A (slow reacting substance of anaphylaxis). These chemical mediators cause contraction of smooth

muscles of the respiratory and intestinal tracts as well as increase vascular permeability.

- *Precipitating factors*—the risk of anaphylaxis is increased if the drugs are given parenterally, family history of allergy, history of asthma and administration of high risk allergens such as penicillin.
- *Onset*—anaphylactic reaction may occur within seconds of drug administration or occur 30 to 40 minutes later. The generalized reaction occurs in four systems cardiovascular, intestinal, respiratory and skin.
- *Skin*—the first sign occurs on skin and it includes urticaria, angioedema, erythema and pruritus.
- *Pulmonary symptoms*—pulmonary symptoms include dyspnea, wheezing and asthma.
- *Gastrointestinal tract*—gastrointestinal tract symptoms like vomiting, cramps and diarrhea occur.
- *Cardiovascular system*—if these are untreated, symptoms of hypotension appear that result from the loss of intravascular fluid. If left untreated, this leads to shock.
- *Prognosis*—the patients with generalized anaphylactic reaction may die from respiratory failure, hypotensive shock or laryngeal edema.

Management

- *Cutaneous rashes*—oral administration or injection of chlorpheniramine, intramuscularly.
- *Angioneurotic edema*—the patient's respiratory distress should be treated immediately with 0.5 ml epinephrine, 1:1000 subcutaneously or 0.2 ml injected slowly intravenously. When immediate danger is passed, then 50 mg of diphenhydramine hydrochloride should be given 4 times daily, until the swelling is diminished.
- *Severe symptoms*—injection hydrocortisone sodium succinate or injection epinephrine.
- *Serum sickness*—It is self-limiting with spontaneous recovery within 1 to 3 weeks. Other treatment is symptomatic with aspirin, for arthralgia and antihistamines, for skin rashes.
- *Anaphylactic reaction* with sudden cardiovascular and respiratory collapse can be treated by following method:-
 - Make the patient to lie down in supine position.
 - Patent airway is to be maintained.
 - Administer 100% O₂.
 - Injection epinephrine 1.1000 IM. It should be given if the blood pressure falls below 60 mm of Hg.
 - For bronchospasm, slowly inject aminophylline 250 mg IV over a period of 10 minutes. Too rapid administration can lead to fatal cardiac arrhythmias.
 - Start IV infusion stat.
 - Monitor pulse, blood pressure and respiration.
 - Hydrocortisone 100 mg IV.

- Antihistaminic should be given.
- Refer to physician and arrange for hospitalization.

Syncope

It is also called as 'simple faint', 'swoon', 'psychogenic syncope', and 'vasodepressor syncope'. It is a transient loss of consciousness due to cerebral anoxia.

Causes

- *Young and poor health person*—it is commonly seen in younger individuals and people having poor health.
- *Precipitating factors*—the other factors are anxiety, fear, and sight of blood, pain, exhaustion, fasting and hot environment. These emotional stresses induce release of increased amount of catecholamine.
- *Cerebral anoxia*—the chief cause is cerebral anoxia or anemia. A blood loss of one liter will cause fainting.

Clinical Features

- *Presyncope*—victim falls gently to the floor, regains consciousness almost immediately or within a short period of time, appears to recover completely.
- *Early signs and symptoms*—the patient complains for warmth in the neck and face. He bathes in beads of cold sweat. He also complains of nausea and feeling vague. Blood pressure at this time is at baseline or slightly lower than baseline. There is also rapid heart rate. Face is pale or has ashen gray skin tone.
- *Late signs and symptoms*—as the process continues papillary dilation, yawning, hyperpnea, coldness in hand and feet, hypotension, bradycardia, visual disturbances, dizziness and loss of consciousness occur.
- *Syncope*—with loss of consciousness, breathing may become irregular, jerky and gasping. The pupils of eye dilate and patient has deathlike appearance. Convulsive movements or muscular twitching of hands, legs or facial muscle. Blood pressure is low and pulse rate is also decrease.
- *Post syncope*—with proper positioning, recovery usually appears rapidly. Patient exhibits pallor, nausea, weakness and sweating, which may persist for few minutes to few hours.

Management

- *Supine position*—when there are signs of fainting he/she should be made to lie down in supine position with legs raised to improve venous return to the heart (**Fig. 33-17**). In case of patient being on the dental chair the back of the chair should be immediately lowered so that the head of the patient is at a lower level than the feet.



Fig. 33-17: Supine position to be maintained in the syncope.

- *Loosening of tight clothing and belt*—tight clothing and belt should be loosened.
- *Maintenance of patent airway*—a patent airway should be maintained. Inhalation of aromatic spirit or application of cold sponge to the face helps in securing reflex stimulation.
- *Oxygen administration*—if cyanosis is developing, 100% oxygen is administered and vital signs are recorded.
- *Ammonia ampule*—an ammonia ampule is crushed and held under the patient's nose for speed recovery.
- *After recovery*—after complete recovery, patient should be slowly brought to semi-reclining, rather than sitting, position.
- *Prevention*—it is usually advisable to treat patients in supine or semi-reclining position. The role of pre-medication before surgery should be considered. Patient who is fasting is more prone to vasovagal attack.

Shock

If the primary shock or syncope is not tackled and allowed to persist, the secondary or true shock appears. Shock can be hemorrhagic, hypovolemic, septic, anaphylactic shock.

Clinical Features

- *Symptoms*—the patient is unconscious with ashen gray face and cold clammy skin.
- *Signs*—mucous membrane is pale, whereas lips, nails, fingertips and lobules of ear are grayish blue. Pulse is weak and thready.
- *Face*—face is expressionless with sunken eyes.
- *Pupil*—pupils are dilated, but react feebly to light.

Management

- *Establishing the cause*—establish the cause of shock like loss of blood, extremely painful stimulus, emotional reasons, toxemia or anaphylaxis and the treatment should be given.
- *Patient position*—put the patient in position with head at lower level than feet.
- *Maintenance of body heat*—maintain the body heat by covering the patient with blanket and keeping a hot water bottle between the thighs.
- *Airway management*—check the patency of airway and control the loss of blood by pressure packs, ligation of vessels or crushing of bone.
- *Restoration of body fluids*—restore the lost body fluids; Ringer's lactate solution should be used to maintain the intravenous line and to restore the volume loss. If hemoglobin is fallen due to toxemia, packed cells are given.
- *Oxygen*—administer 100% of oxygen to the patient for adequate oxygenation of the body tissue.
- *Monitoring of vital signs*—the blood pressure, pulse rate and respiration rate should be constantly monitored to assess signs.
- *Steroids injection*—injection hydrocortisone sodium hemisuccinate 100 mg, dissolved in 5 ml of sterile water is given.
- *Mephentermine injection*—injection mephentermine is given for hypotension.
- *Injection atropine*—if pulse is weak injection atropine is diluted with 5 ml of distilled water and is injected slowly till the radial pulse becomes palpable.
- *Antibiotics*—broad spectrum antibiotics through IV route are given.
- *Adrenaline*—one ampule of 1:1000 dilution of adrenaline in 10 ml of sterile water is given intravenously.
- *Painkiller*—a potent painkiller like narcotic analgesic to gain relief from pain.

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34

Diseases of Bone Manifested in Jaw

4

Introduction

Bone is a dense calcified tissue which is specifically affected by a variety of disease that often causes it to react in a dynamic fashion. These diseases of bone may arise at any age; some are congenital and present at birth, while other develop in early childhood or in young adulthood.

Fibro-osseous Lesions

In it, normal bone is replaced by benign fibrous tissue showing varying amounts of mineralization. The subject of benign fibro-osseous enlargement of the jaws has many facets, the most sinister of which is the possibility that a healthy good looking person may take on a monstrous appearance.

Classification

First Classification

- *Fibro-osseous lesions of medullary bone origin*
 - Fibrous dysplasia
 - Fibro-osteoma
 - Cherubism
 - Juvenile ossifying fibroma
 - Giant cell tumor
 - Jaw lesions in hyperparathyroidism
 - Paget's disease
- *Fibro-osseous lesions of periodontal origin*
 - Periapical cemental dysplasia
 - Florid osseous dysplasia
 - Cemento-ossifying fibroma
 - Cementifying fibroma
 - Ossifying fibroma

Second Classification

Second classification given in 1993 by Waldron reclassified these lesions as follows:

- *Fibrous dysplasia*
- *Reactive (dysplastic) lesions arising in the tooth bearing area*—These are presumably of periodontal ligament origin. Depending on their reference they are divided into three types although they seem to represent the same pathologic process:
 - Periapical cemental dysplasia
 - Focal cemento-osseous dysplasia
 - Florid cemento-osseous dysplasia
- *Fibro-osseous neoplasms*—These are widely designated as cementifying fibroma, ossifying fibroma, or cemento-ossifying fibroma.

Fibro-osseous Lesions of Medullary Bone Origin

Fibrous Dysplasia

It arises from the bone forming mesenchyme in the spongiosa and develops by proliferation of fibrous tissue. Lichtenstein in 1938 coined the term 'fibrous dysplasia'. It is also called as 'fibrocystic disease', 'osteitis fibrosa localisata', 'focal osteitis fibrosa' and 'fibro-osteodystrophy'.

There is no general agreement as to the etiology of lesion. It appears to have no familial, hereditary or congenital basis.

Etiopathogenesis

- *Developmental*—Jaffe and Lichtenstein considered it as a developmental anomaly caused by aberrant activity in the bone forming mesenchymal tissue. Most theories

favor a developmental anomaly because the disease begins in early life and is active during the growth period.

- *Endocrine disturbances*—Sternberg and Joseph considered complex endocrine disturbances with local tissue susceptibility as the cause.
- *Genetic*—fibrous dysplasia results from postzygotic mutation in GNAS I (guanine nucleotide binding protein) gene. If mutation occurs in early embryonic period, the polyostotic type of fibrous dysplasia will occur. If mutation occurs during postnatal life, monostotic type of fibrous dysplasia occurs.

Classification

First classification

- *Monostotic fibrous dysplasia*—in it, only one bone is involved.
- *Polyostotic fibrous dysplasia*—in it, more than one bone is involved
 - *Jaffe type*—fibrous dysplasia involving variable number of bone, accompanied by pigmented lesions of the skin or 'Café-au-Lait' spots.
 - *Albright's syndrome*—a severe form of fibrous dysplasia involving nearly all the bones in the body, accompanied by pigmented lesions of the skin plus endocrine disturbances of various types.

Second according to Stewart

- *Monostotic*—indicates involvement of a single bone.
- *Monomelic*—it refers to the involvement of one extremity and is rarely found.
- *Polyostotic*—many bones involved.
- *Albright's syndrome*

Subclinical fibrous dysplasia

Many a times, an unsuspected lesion of fibrous dysplasia comes to light accidentally on routine radiographic examination, without any clinical evidence of the suspected disease. Such examples are termed as subclinical fibrous dysplasia.

Clinical Features

Monostotic fibrous dysplasia

- *Age and sex*—fibrous dysplasia discovered in young patients, usually in children younger than 10 years affecting both the sexes equally.
- *Sites*—monostotic fibrous dysplasia involves only one bone and presents no extra-skeletal effects, other than occasional pigmented skin lesions. Most frequent sites are ribs, femur, maxilla and mandible.
- *Appearance*—swelling is seen on affected site (**Fig. 34-1**).

Polyostotic fibrous dysplasia (Jaffe's type)

- *Sex*—polyostotic fibrous dysplasia involves multiple bones, with female to male ratio of 3:1.
- *Sites*—the most common site for these are the back, buttocks, thighs, shoulders, chest, neck and face in the mentioned order.
- *Skin lesions*—the skin lesions consist of irregularly pigmented, light brown melanotic spots, described as 'café-au-lait spot'.
- *Symptoms*—recurrent bone pain is the most common presenting skeletal symptom.
- *Skeletal lesions*—skeletal lesions may be unilateral in distribution or may involve nearly all bones of the body. Skeletal lesions become static with the cessation of growth but proliferation may continue, particularly in the polyostotic form.
- *Complication*—spontaneous fracture is a common complication.
- *Signs*—in rare cases, continuous and inexorable extension result in great deformity and blindness.



Fig. 34-1: Monostotic fibrous dysplasia showing swelling on face (Courtesy Dr Parate).

McCune-Albright's syndrome

- *Sex*—Albright's syndrome is exclusively found in females.
- *Features*—Albright's syndrome, in addition show, endocrinal disturbances like precocious puberty, goiter, hyperthyroidism, hyperparathyroidism, Cushing's syndrome and acromegaly.
- *Café au lait spot*—these are coffee with milk color spot. There is irregular flat area of increased skin pigmentation.
- *Symptoms*—vaginal bleeding has been noted.
- *Signs*—secondary sexual characteristics such as pubic and axillary hair and development of breasts are evident by the age of 5 years. It may result in crippling deformities or fracture

- *Precocious puberty*—it is rare in boys and is manifested as gynecomastia.
- *Sites*—long bones are frequently affected.
- *Prognosis*—generally, the active period ceases in adult life. If growth continues the rate of activity is markedly reduced. However, calcification of the fibrous tissue may continue for years. Occasionally, the lesions may remain inactive for several years. For some unknown reasons they may renew the activity.

Oral Manifestations

Monostotic

- *Sites*—maxilla (**Fig. 34-2**) is more commonly affected than mandible, with most changes occurring in the posterior region. Most common area involved is premolar-molar area.
- *Symptoms*—swelling is usually painless but patients may feel discomfort in some cases and while others complain of frank pain.
- *Cortical plates*—enlarging deformities of alveolar process mainly buccal and labial cortical plates.
- *Mandible*—in mandible, it causes protuberant excrescence of the inferior border of mandible.
- *Teeth*—the teeth present in the affected area are either malaligned (**Fig. 34-4**) and tipped or displaced. Dental anomalies such as supernumerary teeth have been reported in connection with the monostotic fibrous dysplasia. The most commonly affected site is maxillary midline and mandibular premolar region. These supernumerary teeth often remain impacted and may affect the eruption of normal teeth.



Fig. 34-2: Swelling seen on right side in maxillary region in case of fibrous dysplasia.

- *Appearance*—there may be unilateral facial swelling, which is slow growing with intact overlying mucosa (**Fig. 34-3**).



Fig. 34-3: Unilateral swelling seen on right side with intact overlying mucosa (Courtesy Dr Bande).



Fig. 34-4: Malalignment of teeth seen in upper arch on right side due to fibrous dysplasia (Courtesy Dr Ashok L).

Craniofacial fibrous dysplasia

- *Sites*—if fibrous dysplasia extends to involve the maxillary sinus, the zygomatic process, floor of orbit and sometimes, it extends toward the base of the skull, known as craniofacial fibrous dysplasia.
- *Symptoms*—it results in severe malocclusion and marked facial deformity. Craniofacial lesions may lead to anosmia (loss of sense of smell), deafness and blindness.
- *Signs*—there may be proptosis of the affected eye.

Polyostotic

- *Appearance*—expansion and deformities of jaws. Asymmetry of facial bones. There is ballooning of jaws, so there is gross enlargement and deformity.
- *Teeth*—the eruption pattern of teeth is disturbed because of loss of support of the developing teeth.
- *Pigmentation*—in some cases, intraoral pigmentation can be seen.

Radiographic Features

Lesions showing predominance of fibrous tissue

- *Early*—radiolucent with ill defined borders. The bony defect may be often unilocular but occasionally bony septa may be apparent creating an impression of multilocular cavity.
- *Margins*—margins may be well defined with a tendency to blend imperceptibly with surrounding normal bone.
- *Granular appearance*—surrounding the margins of the radiolucent area, there may be wider band of increased density, but granular in appearance (**Fig. 34-5**).
- *Lamina dura*—when the lesion involves the apices of teeth there is loss of lamina dura or if retained, it has less density than normal.
- *Teeth*—resorption of roots and destruction of developing teeth.
- *Jaws*—when the lesion comes to the surface, there may be expansion of the jaws.

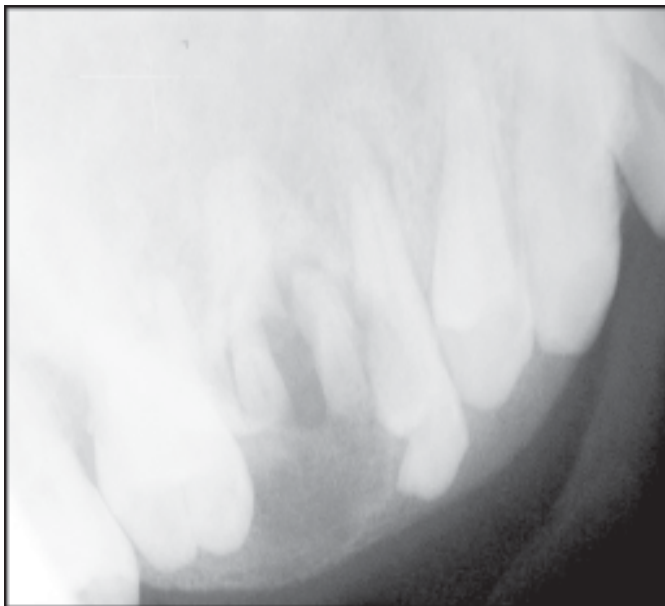


Fig. 34-5: Granular type appearance seen in fibrous dysplasia patient.

Lesions showing mixed radiolucent and radiopaque appearance

- *Appearance*—radiographic appearance of lesions with heterogeneous distribution of fibrous and osseous tissue shows a mixed radiolucent and radiopaque appearance, depending on the maturity of the lesions (**Fig. 34-6**).
- *Granular appearance*—the new bone takes the form of very small opacities of poor density. When they become larger they appear as granular.
- *Maxillary lesion*—it may spread to involve the adjacent bone such as zygoma, sphenoid, occiput and base of skull.

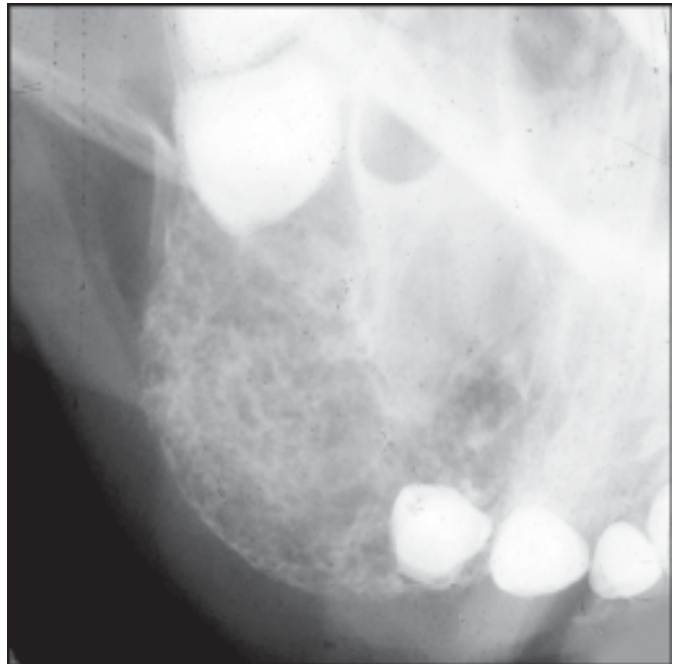


Fig. 34-6: Mixed type lesion seen in fibrous dysplasia,

Mature radiopaque lesions where bone is predominant

- *Stippled*
 - *Orange peel*—the radiograph shows bone of increased density. The normal structure of bone is replaced by a stippled appearance which resembles the ring of orange which is called as 'orange peel'.
 - *Teeth*—tilting and bodily displacement of teeth in the affected area.
 - *Maxillary sinus*—it may obliterate the maxillary sinus.
 - *Thumb print appearance*—when mandible is affected, the vertical depth of mandible is increased. The inferior border of mandible appears as a ribbon like cortex. In some cases, the localized area over the cortex is lost and instead, there is a smooth curved downward projection of the inferior margins of the bone. The appearance resembles a 'thumb print' (**Fig. 34-7**), as if the bone had been soft and pressed upon by the thumb.
 - *Expansion*—bony expansion usually extends to the buccal and distal aspect.
 - *Smoky mottled appearance*—as the lesions mature, dysplastic bony trabeculae increase in size and number and appear like smoky mottled radiopacities.
- *Granular appearance*
 - *Ground glass appearance*—another characteristic appearance of fibrous dysplasia is ground glass appearance, also termed as granular. Apart from the appearance everything else is similar as in stippled type. It may demonstrate areas of whorled amorphous partially calcified materials that are well circumscribed.

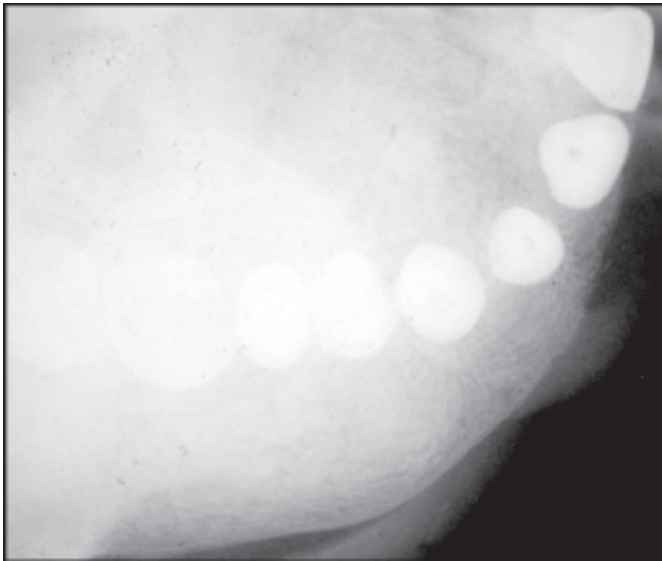


Fig. 34-7: Thumb print appearance seen in fibrous dysplasia.

- *Dense structureless, homogeneous*—in this, the lesion shows a dense structureless, homogeneous appearance (Fig. 34-8). Any part of the joint may be affected. The commonest site is the base of the skull with involvement of the maxilla and nasal bones. Encroachment or obliteration of the antrum and spread into the adjacent bone including the base of skull take place.
- *CT features*—CT is useful to evaluate the extent of facial lesions; especially orbital involvement. The CT characteristics of the fibrous dysplasia include expansion of the involved bones in a heterogeneous pattern with scattered or confluent islands of bone formation (Fig. 34-9). CT attenuation (density) values can reach 34 to 513 Hounsfield units or more particularly in dense lesions. The variation in the values reflects the complex, and often non-uniform, distribution of primitive bony trabeculae.

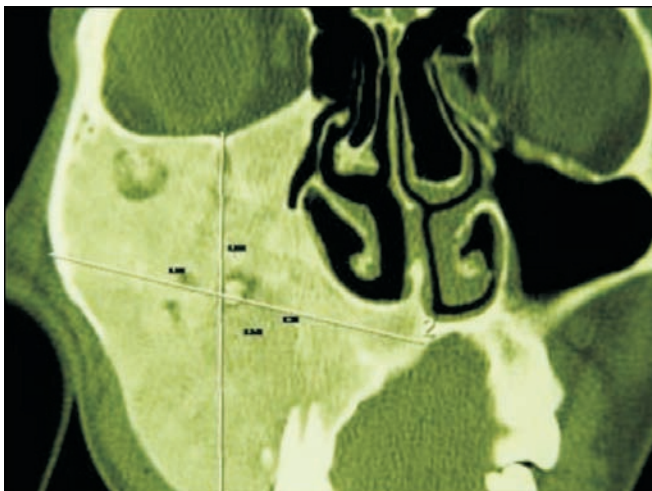


Fig. 34-8: Homogeneous pattern of fibrous dysplasia seen on computed tomography (Courtesy Dr Iswar).

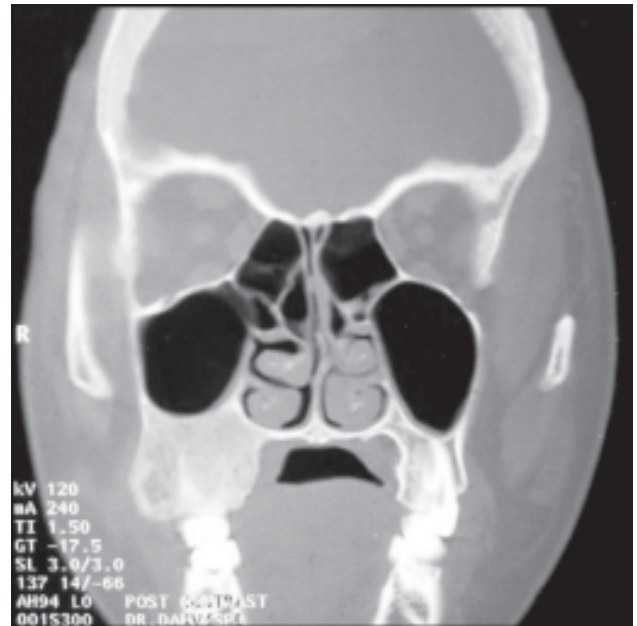


Fig. 34-9: Confluent pattern seen on CT scan on right side in case of fibrous dysplasia.

Obisesan et al classified the lesions of fibrous dysplasia radiographically into 6 types.

- *'Peau d' orange' or orange peel*—in this type, there are alternating areas of granular density and lucency giving a radiographic appearance resembling the ring of orange.
- *Whorled plaque like type*—in this type, the matrix of the well circumscribed lesion is composed of plaques of amorphous material of intermediate radiodensity, which on close examination are seen to be arranged in whorled onion peel appearance.
- *Diffuse sclerotic type*—the lesions of this show as homogeneous dense area, which gradually merges with the normal bone.
- *Cyst like type*—in this type, the lesions are radiolucent. It is unilocular or multilocular, more often multilocular with well defined margins.
- *Pagetoid type*—in this type of lesions, the affected area of bone markedly expands and shows alternating areas of radiopacities and lucency, as those seen in Paget's disease of bone.
- *Chalky type*—it manifests itself as a well circumscribed lesion consisting of an amorphous dense radiopaque material.

Craniofacial fibrous dysplasia

- *Density*—radiograph reveals the presence of a marked density which encroaches to a variable degree upon the orbit and antral cavity.
- *Granular type*—changes in the base of the skull is of granular type and structureless, so that affected portion of the bone is thickened and of greater density.

- *Frontal bone*—the frontal bone is also thickened with homogeneous or variegated type of density.
- *Nasal septum*—the nasal septum is grossly thickened, dense and curved, so that it represents the gross caricature of the letter S.

Diagnosis

- *Clinical diagnosis*—painless swelling seen in the maxillary region with intact mucosa. Café au lait spots are seen.
- *Radiological features*—ground glass appearance, granular appearance is seen.
- *Laboratory diagnosis*—when the polyostotic lesions are numerous and active, the serum alkaline phosphatase levels may be elevated in 50% of the cases. Biopsy shows bone made up of proliferating fibroblast in a compact stroma of interlacing collagen fibers. Irregular trabeculae of bone are scattered throughout the lesion, with no definite pattern of arrangement. Some of these trabeculae are C-shaped and described as Chinese character shaped.

Differential Diagnosis

Lesions are likely to be confused in the osteolytic stage of fibrous dysplasia

- *Central giant cell granuloma*—it has got faint wispy trabeculae coursing through it whereas internal calcifications may occur in fibrous dysplasia and appear to be stippled and granular appearance.
- *Traumatic bone cyst*—there is no cortical bulging and displacement of teeth.
- *Dental cyst*—it has a thin, well defined cortex which is smooth, while in fibrous dysplasia, cortex tends to be wider and more granular in appearance.
- *Aneurysmal bone cyst*—there is hemorrhagic aspirate.
- *Chronic osteitis*—it is always associated with the roots of pulp less teeth.
- *Chronic osteomyelitis*—it manifests itself in older age group (30-80 years) as compared to fibrous dysplasia (10-20 years). In addition, patients with osteomyelitis will give a history of trauma, fracture or any debilitating systemic disease.
- *Peripheral and central squamous cell carcinoma*—it also occurs in older age group and shows a predilection for mandible. Fibrous dysplasia grows by slow expansion whereas central and peripheral squamous carcinoma spreads rapidly.
- *Metastatic tumor*—seen in older age groups and shows a predilection for premolar-molar region of mandible.
- *Reticular cell sarcoma and Ewing's sarcoma* are rarely seen in maxilla.

The mottle type of fibrous dysplasia

- *Lymphoma of bone*—is rare and is poorly defined. The radiographic pattern is irregular and bizarre. In fibrous dysplasia smooth, well contoured external bony borders are always maintained.
- *Chondrosarcoma*—is an uncommon malignant tumor of the jaws which is often painful and affects a much older age group than fibrous dysplasia.
- *Osteoblastic metastatic carcinoma*—seldom shows a monotonous pattern as in fibrous dysplasia. A history of either symptoms or treatment for a primary tumor elsewhere will be elicited during history taking. Osteoblastic metastatic carcinoma is found in older age group.
- *Osteosarcoma*—its appearance is disorderly with sunburst pattern, codmans triangle and asymmetrical band like widening of the periodontal ligament.
- *Paget's disease*—classically, Paget's disease simultaneously affects several bone of the skeleton. It is a disease of the later age group; the serum alkaline phosphatase level is elevated in Paget's disease whereas in fibrous dysplasia, it is within normal limit. Rare in adolescents and young adults. Bilateral involvement.
- *Cementifying and ossifying fibroma*—it exhibits a similar mottled appearance to that seen in fibrous dysplasia. The following differences are recognized.
 - *Shape*—cementifying and ossifying fibroma as predominantly rounded while those of fibrous dysplasia are more rectangular.
 - *Jaw expansion*—jaw expansion caused by cementifying and ossifying fibroma is usually nodular or dome shaped whereas in fibrous dysplasia it is usually the elongated fusiform type.
 - *Margins* of fibrous dysplasia are indistinct, blending imperceptibly with normal bone while in cementifying and ossifying fibroma, the margins are sharply defined.
 - *Predominance in jaws*—approximately 70% of cementifying and ossifying fibroma occur in mandible while fibrous dysplasia shows a slight predilection for maxilla.
 - *Predominant age*—the age range for cementifying and ossifying fibroma is from 7-58 years. The majority of active cases of fibrous dysplasia are found in patients under the age of 20 years.
- *Chronic osteomyelitis*—in it, radiolucent-radiopaque appearance can mimic the mottled appearance of fibrous dysplasia. Generally, when purulent discharge is present, the diagnosis is of chronic osteomyelitis.

Mature phase

- *Paget's disease and giant cell lesion of hyperparathyroidism*—a solitary painless fusiform enlargement, which is firm smooth, covered by normal mucosa and has a

radiopaque ground glass appearance, occur in the jaw bones of relatively young person is almost certainly fibrous dysplasia. Paget's disease and giant cell lesions of hyperthyroidism can produce ground glass appearance but the overall effect is rarefaction and not radiopacities.

Management

- *Surgical*—surgical removal of the lesion should be carried out.
- *Osseous contouring*—it is necessary for correcting the deformity for esthetics or pre-esthetic purposes.

Juvenile Ossifying Fibroma

It is also called as 'young ossifying fibroma', 'juvenile aggressive ossifying fibroma', and 'trabecular desmoosteoblastoma'. This name has been given as this occurs in younger age group.

Classification

Slootweg and colleagues have separated the lesions into two distinct groups:

- *Juvenile ossifying fibroma WHO type*—it occurs predominantly in the maxilla and mandible. Here, the designation of juvenile ossifying fibroma should be restricted exclusively to those lesions whose histomorphology is consistent with that described in the 1992 WHO monograph on jaw lesions. This trabecular variant has strands of immature cellular osteoid within the lesion and usually occurs in childhood with a slight maxillary predilection.
- *Juvenile ossifying fibroma with psammoma like ossicles*—it occurs in the paranasal sinuses and extragnathic bones. The psammomatoid variant has small spherical ossicles surrounded by osteoid rims within the lesion. It occurs over a wider age range than the trabecular variant and usually affects the orbit or paranasal sinuses.

Clinical Features

- *Age*—it is seen under the age of 15 years.
- *Site*—no site predilection and with equal frequency in both the jaws. Although there is report that this is more commonly found in maxillary region.
- *Symptoms*—the presenting clinical symptom is of swelling (Fig. 34-10).
- *Effect on surrounding structure*—neoplasm can impinge on neighboring structure. Due to impingement, patient may noticed nasal obstruction, exophthalmos and proptosis. In some cases, permanent blindness may occur.
- *Intracranial extension*—in some cases, meningitis may arise due to intracranial extension.



Fig. 34-10: Swelling seen in maxillary region due to ossifying fibroma.

Radiographic Features

- *Internal structure*—it may appear as unilocular or multilocular lesion.
- *Appearance*—it may be radiolucent or show mixed appearance containing radiopaque foci (Fig. 34-11).
- *Margin*—it has got distinct radiopaque border.



Fig. 34-11: Juvenile ossifying fibroma showing mixed radiopaque radiolucent lesion (Courtesy Dr Bande).

Diagnosis

- *Clinical diagnosis*—it is not possible to make clinical diagnosis.
- *Radiological diagnosis*—mixed lesion in young patient will give clue to the diagnosis.
- *Laboratory diagnosis*—biopsy shows non-encapsulated lesion. There are also myxomatous foci, nuclear crowding, and multinucleated osteoclasts.

Differential Diagnosis

- *Ossifying fibroma*—juvenile ossifying fibroma occurs at a far lower mean age (8.5 years) than ossifying fibroma (26.4 years). Ossifying fibroma contains lamellar bone and cementicles as well as smoothly contoured cells, poor curvilinear trabeculae, features that are absent in juvenile ossifying fibroma.

Management

- *Excision*—complete local excision of tumor is the treatment of choice.

Cherubism

It is also known as 'familial fibrous dysplasia of the jaws', 'disseminated juvenile fibrous dysplasia', 'familial multilocular cystic disease of the jaws' and 'Hereditary fibrous dysplasia of the jaws'. Above terms of cherubism are related to fibrous dysplasia but there is no similarity between fibrous dysplasia and cherubism so, this term should be avoided.

The clinical entity was first described by Jones in 1933 who coined the term 'Cherubism' reflecting the characteristic of chubby facial appearance similar to plump cheeked little angle (cherubs) seen in Renaissance painting. It is autosomal dominant inherited fibro-osseous disease that affects only the jaws causing bony expansion.

Classification

It depends on the severity and location of the lesion and the extent to which jaws are affected.

- *Grade I*—the fibro-osseous expansion tends to be bilateral and symmetrical. It is primarily in the ramus of the mandible.
- *Grade II*—in more severe cases, the ramus and the body of the mandible are involved resulting in congenital absence of the third and occasionally the second mandibular molar teeth. In this group, the tuberosity region of the maxillae is also affected.
- *Grade III*—in these cases, the lesions affect the mandible and maxilla entirely and may result in considerable facial deformities.

Etiology

- *Developmental defect*—anomalous development of dental structures with disturbance in the development of bone forming mesenchyme may lead to this condition.
- *Hormonal*—cherubism can be present in latent hyperparathyroidism and hormone dependent benign neoplasm.
- *Other*—other factors which are responsible for cherubism are trauma and an aberration in ossification.

Clinical Features

- *Age and sex*—early childhood between the ages of 2 to 4 years males are affected about twice as frequently as females.
- *Sites*—it shows predilection for angle of mandible bilaterally and occasionally posterior maxilla.
- *Appearance*—in the rapidly increasing stage, the child assumes a *chubby, cherubic facial appearance* (Fig. 34-12), especially if combined with involvement of the orbital floor with upward displacement of the globe and exposure of the scleral rims. This appearance results due to bilateral involvement of posterior mandible.



Fig. 34-12: Cherubic appearance of the child in cherubism.

- *Symptoms*—the patient may have difficulty in speech, deglutition, mastication, respiration and limited jaw movement.
- *Signs*—swelling is firm and hard on palpation (Fig. 34-13). Overlying mucosa is intact and non-painful.



Fig. 34-13: Firm swelling seen in the jaw of patient of cherubism.

- *Jaws*—bilateral enlargement of mandible in this condition produces full, round lower face. Bilateral enlargement of maxilla gradually follows.
- *Eye raised to heaven*—pulling or stretching of skin of the cheek, depresses the lower eyelid, exposing a thin line of sclera and resulting in the so called “eyes raised to heaven” look.
- *Orbital floor*—tumor encroachment on the orbital floor often causes partial obliteration of the palatal vault with resulting ‘V’ shaped cleft.
- *Alveolar process*—the alveolar process are so wide, as to occupy almost the whole of the roof of the mouth; the actual palate being reduced to a narrow fissure between the two approximating alveolar process.
- *Lymph nodes*—there may be enlargement of sub-mandibular lymph nodes.
- *Progress*—there is rapid increase in size up to 7-8 years of age, after which the lesions become static or progress very slowly until puberty. After puberty the maxillary lesions tend to regress. The mandibular lesions progress slowly up to the age of 20 years and then regress; the facial appearance almost returns to normal in the 4th and 5th decades of life.
- *Teeth*—the fibrous replacement of bone displaces the deciduous dentition. The primary teeth may be irregularly spaced and some may be absent. There is premature loss of primary teeth. The developing permanent teeth are affected, giving rise to displaced unerupted or absent teeth along with malocclusion.

Radiographic Features

- *Radiodensity*—cyst like radiolucency of mandible, bilaterally symmetrical which is up to several centimeters in diameter.
- *Progress*—initiation of bone destruction near the angle of the mandible with later expansion of lesions posteriorly into ramus and anteriorly into the mandibular body.
- *Appearance*—it appears as a classic multilocular cavity due to internal radiopaque septa, which tends to coalesce as they enlarge. On posteroanterior views, teeth are seen to be hanging in air.
- *Margins*—they are well defined, well corticated and smooth around most of the radiolucency.
- *Effect on surrounding structure*—there is also expansion of buccal and lingual cortical plates (Fig. 34-14). In mandible, the inferior alveolar canal may be displaced and the lesion may occupy alveolar process, the angle and the ramus. The thin cortex may eventually disappear. Maxillary lesions enlarge at the expense of maxillary sinus.
- *Effect on teeth*—displacement of numerous teeth but prior to enamel calcification. Erupted deciduous teeth in the

area of bone involvement shed prematurely. Few posterior teeth may be missing due to early developing expanding masses which destroy the buds and incipient follicle.



Fig. 34-14: CT scan of patient of cherubism showing bilateral swelling in mandibular posterior region (Courtesy Dr Iswar).

Diagnosis

- *Clinical diagnosis*—typical cherubic appearance of child with bilateral swelling in the jaw
- *Radiological diagnosis*—cyst like radiolucency present bilaterally in the mandible
- *Laboratory diagnosis*—in active cases, serum alkaline levels may be raised. Biopsy shows large multinucleated giant cell in a loose delicate fibrillar connective tissue stroma.

Differential Diagnosis

- *Fibrous dysplasia*—cherubism is bilateral.
- *Giant cell granuloma*—it occurs frequently in the anterior segment of mandible in contrast to cherubic lesions, which are seen in the posterior part of mandible.
- *ABC*—the lesions of ABC are tender, whereas cherubic lesions are painless.
- *Central hemangioma*—there is localized gingival bleeding and pumping tooth syndrome.
- *Giant cell lesions of hyperthyroidism*—are not bilateral and they can be differentiated on abnormal blood chemistry levels.
- *Metastatic tumors*—it is seen in older age group as compared to cherubism and also there will be signs and symptoms or history of primary tumor elsewhere in the body.
- *Ameloblastoma*—it is seen unilaterally and frequently in an older age group than that of cherubism
- *Odontogenic myxoma*—history of missing tooth, as it is of developmental origin.

- *Nevoid basal cell carcinoma*—no facial swelling is seen in cherubism. There are characteristic cutaneous abnormalities or rib anomalies seen on chest radiograph.
- *Multiple dentigerous cyst*—impacted teeth present.

Management

- *Surgical contouring*—surgical procedures should be delayed, as long as possible, as the cystic lesion defect usually becomes static and regresses during adulthood. Occasionally, surgical contouring of the lesions is necessary to improve esthetics and in case of active growth.
- *Homogeneous bone grafts*—in cases of extensive involvement, homogeneous bone grafts can be given to prevent pathological fractures of mandible.
- *Orthodontic care*—orthodontic care may be required to ensure proper alignment of the teeth.
- *Calcitonin*—this therapy is under trial nowadays, awaiting results of some study.

Central Giant Cell Granuloma

It is a non-neoplastic bone disease reactive to some unknown stimulus. It was first described in the jaws by Warren 1837. It has been called as *osteoclastoma*, *myeloid sarcoma*, *chronic hemorrhagic osteomyelitis* and *giant cell reparative granuloma* initially. But nowadays the term reparative is dropped and it is called as *giant cell granuloma* only.

Types

- *Non-aggressive*—it exhibits slow growing benign behavior.
- *Aggressive*—it shows typical features of rapidly growing, destructive lesion.

Clinical Features

- *Age*—lesion of adolescent and young adult with 60% cases in younger than 20 years and 74% cases in younger than 30 years.
- *Sites*—mandible is twice as frequently involved than maxilla, with anterior half showing greatest incidence with fairly high percentage crossing the symphysis. The commonest site being the anterior and bicuspid region in mandible and the canine fossa and ethmoid region in maxilla.
- *Symptoms*—the earliest sign of the lesion may be expansion of bone with premature loosening and shedding of deciduous teeth. There is jaw swelling associated with facial asymmetry. Usually painless, but local discomfort may be noted.

- *Signs*—palpation may elicit tenderness. Growth is slow.
- *Teeth*—teeth in the area may become mobile but maintain their vitality, until they are exfoliated.
- *Prognosis*—recurrence rate of 12% for lesions under 2 cm and 37% recurrence for lesions over 2cm in size.

Radiographic Features

- *Radiodensity*—solitary unilocular (**Fig. 34-15**) or multilocular radiolucency.

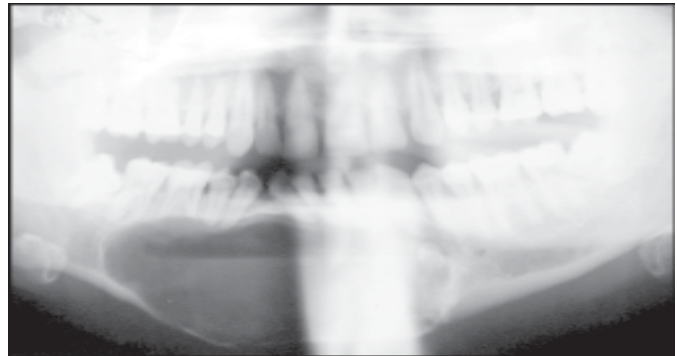


Fig. 34-15: Unilocular radiolucency seen in mandibular anterior region.

- *Location*—it may occupy whole of the mandibular body and may extend past the midline to the opposite side (**Fig. 34-16**).
- *Appearance*—as it grows, it causes bossing of buccal cortex i.e. uneven, variable bulging or undulation of the cortical contour.
- *Margins*—borders may be smooth, undulating, moderately well defined and moderately well corticated.
- *Honeycomb appearance*—it shows bony trabeculation within their contour and tends to be mildly wavy on close inspection. It may have wispy delicate quality. Sometimes, they may have honeycomb appearance.



Fig. 34-16: Radiolucency seen in mandible crossing the midline.

- *Effect on surrounding structures*—when they involve maxillary bone, they may erode or expand bone. Displacement of adjacent teeth, tooth buds and resorption may occur.

Diagnosis

- *Clinical diagnosis*—expansive swelling in the anterior region of mandible may suspect central giant cell granuloma.
- *Radiological diagnosis*—radiolucency crossing the midline is typical of central giant cell granuloma (**Fig. 34-17**).
- *Laboratory diagnosis*—biopsy shows loose fibrillar connective tissue stroma with many interspersed proliferating fibroblast and small capillaries. Multinucleated giant cells are prominent.

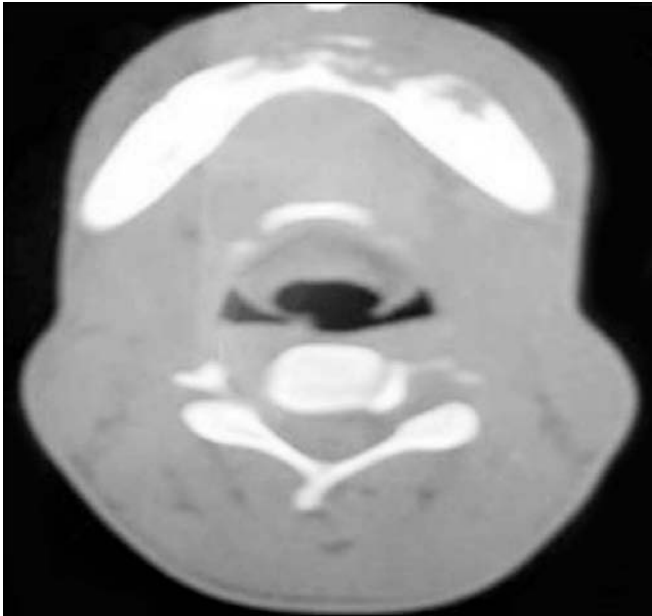


Fig. 34-17: CT scan of central giant cell granuloma showing destructive lesion in mandibular anterior region

Differential Diagnosis

- *Ameloblastoma*—it is uncommon in a younger age range, which is most susceptible to giant cell granuloma. Seen in posterior mandible in contrast to giant cell granuloma which occurs anterior to the first molar. Ameloblastoma demonstrates internal, hard curved arch like septa whereas giant cell granuloma has lighter wispy septa. Ameloblastoma is usually multiloculated.
- *Aneurysmal bone cyst*—it does not occur in anterior segment of mandible. Aspiration produces blood.
- *Odontogenic myxoma*—multiloculated and typical honey-comb appearance. Missing or impacted tooth is usually a finding.
- *Giant cell tumor*—it is described in **Table 34.1**
- *Traumatic bone cyst*—no bodily movement of teeth is present. No expansion of overlying bone cortex.
- *Fluid filled odontogenic cyst*—internal septa are not present. No bony spicules protrude from radiographic margins. Regular smooth bony expansion of cortical plates while scalloped, undulating bulging of cortex.
- *Brown tumors of hyperthyroidism*—serum calcium levels are elevated.
- *Cherubism*—it is bilateral in the posterior part of mandible and there is history of familial involvement. It does not cross the midline.
- *Metastatic tumors*—it is seen in older age groups and also predominant lesions are multilocular with history and symptoms of primary tumor in addition to local lesions.
- *Central hemangioma*—it shows localized bleeding around the necks of teeth and also the pumping tooth syndrome.
- *Post extraction socket, surgical defect and residual cyst*—it can mimic unilocular appearance of central giant cell granuloma. Patients with surgical defect will give a history of previous surgery and history of extraction in residual cyst and post extraction socket.

Table 34.1: Difference between giant cell tumor and giant cell granuloma

<i>Giant cell tumor</i>	<i>Central giant cell granuloma</i>
It is more common in 3rd and 4th decades and unusual in patient below 20 years of age common in males	It occurs mainly in children and young adults often in females
Common in long bone like femur, tibia and rare in jaw	Most common in mandible, appearing anterior to 1st molar tooth bearing area
Microscopically tumor giant cell contains more nuclei and has larger dimension	It contains fewer nuclei and smaller dimension
Even distribution of giant cells throughout the tumor, in every field	In it giant cells are unevenly distributed
It is not preceded by trauma	It is preceded by trauma
New bone formation is absent	Foci of osteoid and new bone are frequently present

Management

- **Curettage**—thorough curettage of the lesion should be carried out.
- **Corticosteroids**—weekly injection of corticosteroids directly into tumor with triamcinolone acetonide can give dramatic results. This should be given weekly.
- **Calcitonin**—systemic administration of salmon calcitonin can lead to resolution of lesion. It should be given daily for 12 months as intradermal injection or nasal spray.
- **Interferon alfa**—this is also given in some cases of central giant cell granuloma.
- **Surgical**—partial resection should be done in case of aggressive tumor.

Paget's Disease

It is also called as '*osteitis deformans*'. It was discovered in 1877 by Sir James Paget. It fondly refers to as '*collage of matrix madness*'. There is abnormal resorption and apposition of bone in one or more bones. The disease is initiated by an intense wave of osteolytic activity with resorption of normal bone resulting in irregularly shaped resorption cavity followed by vigorous osteoblastic activity forming woven bone after variable period.

Etiology

- **Inflammatory**—initially it is thought to be inflammatory disease, but later on this cause is ruled out.
- **Circulatory disturbance**—the bone in Paget's disease is excessively vascular and it has been suggested that vessels are similar to arteriovenous aneurysms.
- **Slow virus theory**—slow viruses are those viruses which take a long time for incubation. The possibility of an infective, viral Etiology for Paget's disease is suggested by ultra-structural demonstration of intra-nuclear inclusions in abnormal osteoclast.
- **Genetic and environmental factors**—genetic and environmental factors appear to be important. It is transmitted as autosomal dominant trait with genetic heterogeneity.
- **Connective tissue factors**—it may be a disorder of connective tissue biosynthesis.
- **Others**—vasculitis, trauma, hormonal imbalance, degenerative neurologic disorders.

Clinical Features

- **Geographical prevalence**—it is seen most frequently in Britain and less frequently in North America and Western Europe.
- **Age and sex**—predominantly in patients over 40 years of age with a slight predilection for men. Male to female ratio is 2:1.

- **Sites**—it is prone to occur in the axial skeleton especially the skull, femur, sacrum and pelvis. It is usually polyostotic.
- **Symptoms**
- **Skull enlargement**—first complaint is that patient needs to buy a hat of larger size because of skull enlargement.
- **Pain**—bone pain is a consistent symptom and most often directed towards weight bearing areas. The patients may have ill defined neuralgic pain as a result of restriction of foramina and canals, which occurs due to pressure created by mass on structures passing through the foramina.
- **Deafness**—deafness due to involvement of the petrous portion of temporal bone with compression of cochlear nerve in the foramen.
- **Neurological symptoms**—there may be mental disturbance, and dizziness.
- **Signs**—bowing of legs, curvature of spine and enlargement of skull. The involved bones are warm to touch because of increased vascularity and are prone to fracture.
- **Simian appearance (likes an apes or monkey)**—due to grotesque facial appearance along with waddling gait and short stature patient has got simian appearance.
- **Waddling (short step) gait**—broadening and flattening of the chest and spinal curvature. The patient assumes waddling gait.
- **Complication**—skeletal deformities, fracture of limbs, compression of spine, and occurrence of giant cell lesion. Another complication includes renal calculi, salivary calculi and hypercalcemia.

Oral Manifestations

- **Sites**—maxilla is involved three times more commonly than mandible. It is bilaterally symmetrical in the involved jaw.
- **Symptoms**—movement and migration of affected teeth occurs. Due to migration patient may noticed malocclusion. In case of edentulous patient as alveolar ridge becomes enlarged patient complaint of poor fit of denture.
- **Signs**—increase in alveolar width associated with flattening of palate when maxilla is involved (**Fig. 34-18**).
- **Appearance**—as the disease progresses, the mouth may remain open exposing the teeth as the lips are too small to cover the enlarged jaws.
- **Extraction sites**—extraction sites heal slowly and incidences of osteomyelitis are higher. Extraction may be further complicated by excessive bleeding from highly vascular abnormal bones in the lytic phase of disease.
- **Complications**—osteogenic sarcoma, osteomyelitis, pathological fracture and facial paralysis. Sarcoma is



Fig. 34-18: Patient of fibrous dysplasia showing increases width of alveolar ridge (Courtesy Dr Bande).

suspected in cases that experience a marked increase in intensity of bone pain or other symptoms and show marked increased in alkaline phosphatase.

- **Prognosis**—it is poor in patients showing development of osteogenic sarcoma in Paget's disease.

Radiographic Features

Early radiolucent stage

- **Appearance**—inferior cortex of mandible may appear osteoporotic and possess a laminated structure.
- **Trabecular arrangement**—bone pattern in which trabeculae though reduced in number, run linearly in the direction of length of bone and have few intersections between them. This appearance is most commonly seen posterior to bicuspid. In the anterior region, the bony trabeculae are coarse and relatively straighter than normal, but they intersect producing bone spaces that are larger than normal. Coarse and sparse trabeculae, sometimes, tend to converge towards the midline of mandible which is highly suggestive of Paget's disease. In this stage, inferior border of the mandible may appear osteoporotic and have a laminated structure.
- **Effect on root**—root resorption is common.
- **Osteoporosis circumscripta**—in the skull, early lytic lesion may be seen as discrete radiolucent areas termed as *osteoporosis circumscripta*. The margins are somewhat irregular. There is appearance of denser bone around the radiolucency.
- **Lincoln's sign or black beard**—this sign is present in bone Scintigraphy. Bone scan may demonstrate marked uptake throughout the entire mandible. This is called as Lincoln's sign or black beard.

Granular or ground glass appearance

- **Radiopaque patches**—there are rounded radiopaque patches of abnormal bone of greater density within the

radiolucent bone, within which it is not possible to see any actual bone structure.

- **Size**—these densities may be a centimeter large or only a few millimeters in diameter.

A dense, more radiopaque stage

- **Cotton wool appearance**—in later stages, rounded radiopaque patches of abnormal bone are often seen giving an impression of cotton wool. As the fully opacified area becomes more numerous and enlarged, they tend to coalesce. The bone is denser and appears whiter on the radiograph.
- **Enlargement of bone**—it enlarges the affected bone and it may be four times than their normal thickness on lateral radiographs. In some cases there is irregular enlargement of alveolar processes which become prominent and bulge.
- **Prognathism of mandible**—due to deposition of bone in mandible, it may appear prognathic.
- **Effect of maxillary sinus**—lesions in maxillary area may encroach the maxillary sinus and obliterate it.
- **Hypercementosis**—it may be produced on one or more teeth (**Fig. 34-19**).
- **Ankylosis of teeth**—it may obliterate the areas of lamina dura and periodontal ligament space around both, normal and hypercementosed roots resulting in ankylosis of teeth.
- **Osteogenic sarcoma**—development of osteogenic sarcoma may produce frank dissolution or destruction of bone. In it, there is loss of continuity of the margins on the one side of the lesion which is suggestive of the destruction. Also in some cases, soft tissue shadow is seen.

Diagnosis

- **Clinical diagnosis**—in this simian appearance, enlargement of skull, bowing of leg and bone pain may give clue to the diagnosis.

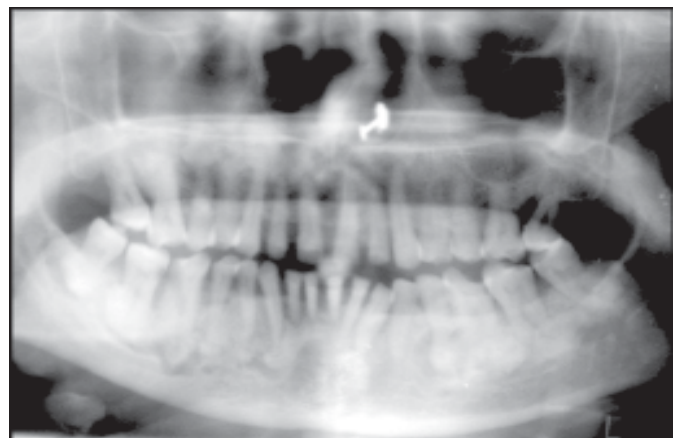


Fig. 34-19: Hypercementosis of teeth in the mandibular teeth (Courtesy Dr Bhaskar Patle).

- *Radiological diagnosis*—cotton wool appearance, Lincoln sign, and osteoporosis circumscripta are diagnostic features of Paget's disease.
- *Laboratory diagnosis*—serum alkaline phosphatase level is increased, occasionally, attaining the level of 200 or more KA units. Serum calcium and serum phosphorus levels are within normal limits. Urinary hydroxyproline levels increase from normal levels of 440 mg/24 hours to 1 gm/24 hours. Histologically there is appearance of mosaic bone.

Differential Diagnosis

Early stage—radiolucent appearance

- *Giant cell lesions of hyperparathyroidism*—in Paget's disease, there is an increase in serum alkaline phosphatase only whereas in Giant cell lesions of hyperparathyroidism there is elevation of serum alkaline phosphatase as well as serum calcium levels with decrease in serum phosphate levels. Also in case of hyperparathyroidism, there is overall radiolucency.
- *Osteoporosis*—in early stages, it may be confused with Paget's disease but if the bony area in question is pathologically enlarged and if straight linear trabeculae are seen in the affected bone parallel to the long axis, osteoporosis is ruled out. Blood chemistry is normal.
- *Osteomalacia*—pseudofractures are common in cases of osteomalacia. Serum calcium and serum phosphorus levels are decreased in osteomalacia whereas in Paget's disease, they are normal.
- *Multiple myeloma*—it causes painful enlargement of the jaws and shows typical radiolucent punched out lesions on the skull radiograph. Positive test for Bence Jones proteins.

Second stage—mixed radiolucent radiopaque appearance

- *Osteogenic sarcoma*—it occurs in a younger age group (10-40) than that of Paget's disease and it shows a variety of radiographic appearance i.e. sunburst and Codman's triangle.
- *Cementifying and ossifying fibroma*—it is predominately seen in the younger age group and they show well defined margins in contrast to that of Paget's disease which is diffuse.
- *Fibrous dysplasia*—age difference, Paget's disease spreads more diffusely, seen bilaterally, cotton wool appearance. Maxillary sinus is reduced in size in the case of fibrous dysplasia but in the case of Paget's disease, it fails to reduce the air space.
- *Osteoblastic metastatic carcinoma*—it will give a history of parent tumor or show signs and symptoms of primary tumor.
- *Ossifying sub-periosteal hematoma*—it can be excluded if the patient gives a recent history of trauma and in addition it is seen in patients less than 15 years of age.

- *Ossifying post surgical defect*—it will give a history of surgery.
- *Chronic osteomyelitis*—it is rare in maxilla.
- *Chondroma and chondrosarcoma*—it produces pain, which is not a feature of Paget's disease.

Advanced stage—purely radiopaque

- *Florid osseous dysplasia*—it is confined to jaw bone and hypercementosis is present.
- *Osteosclerosis*—small and confined to jaw bone.
- *Tori*—same as above.
- *Osteoma*—same as above.

Management

- *Medical*
 - *Calcitonin*—a parathyroid hormone antagonist produced by the thyroid gland, suppresses bone resorption and also relieves pain and decrease serum alkaline.
 - *Sodium phosphate*—it retards bone resorption.
 - *Bisphosphonates*—they have also been used, since they inhibit bone resorption as well as mineralization. Bisphosphonates which are used are etidronate, pamidronate, alendronate, tiludronate or risedronate.
 - *Mithramycin*—it is considered as a second line agent due to its toxicity; is cytotoxic to osteoclasts.
 - *Plicamycin*—it inhibits osteoclastic activity
- *Surgery*—surgical approach for esthetics correction should be carried out.
- *Radiation*—in some cases radiation therapy can be useful.

Giant Cell Lesions of Hyperparathyroidism

It is discussed in the Chapter 36: Endocrine Disorders.

Fibro-osseous Lesions of Periodontal Origin

Periapical Cemental Dysplasia

It is described in Chapter 14: Odontogenic Tumor of Jaw.

Florid Osseous Dysplasia

It is also called as 'chronic sclerosing osteomyelitis', 'sclerosing osteitis', 'multiple enostosis' and 'sclerotic cemental masses'. It is considered to be a widespread form of periapical dysplasia. It is derived from the cells in or near the periodontal ligament space.

Robinson defined it as an abnormal reaction of bone to irritation or stimulation and the term florid osseous dysplasia includes chronic diffuse sclerosing osteomyelitis

and sclerotic cemental masses. It is inherited as an autosomal dominant trait.

In florid osseous dysplasia, the normal bone is replaced by fibrous tissue and mineralized structure like cementum, bone or both.

Clinical Features

- **Age and sex**—females are exclusively affected. Most common age is middle age, with a mean age 42 years with predilection for blacks.
- **Sites**—the lesion is restricted to the jaw bone with mandible being most commonly affected.
- **Symptoms**—there is a painless expansion of the alveolar process of mandible. Patients may complain of intermittent poorly localized pain in the affected bone area, with or without an associated bony swelling.
- **Signs**—if the lesion secondarily infected features of osteomyelitis may develop. Mucosal ulceration with fistulous tract may be present. Teeth in the involved bone are vital.

Radiographic Features

- **Location**—lesions of florid osseous dysplasia present bilaterally in both the jaws. Lesions occur above the inferior alveolar nerve canal. It involves all four quadrants.
- **Radiodensity**—it varies from an equal mixture of radiolucent and radiopaque region to almost complete radiopaque.
- **Size**—individual lesions do not exceed 2-3 cm in diameter. The lesions may extend into the mandibular ramus or into the maxillary sinus.
- **Margins**—margins are fairly regular and well defined. Each lesion is surrounded by a radiolucent capsule and a cortical rim.
- **Radiolucent stage**—in this stage, well defined radiolucent area are superimposed over the apical area of adjacent tooth.
- **Mixed stage**—in this stage radiolucent cavity, partially filled with one or more dense radiopaque masses. As lesions mature, the radiopacities increase. This stage also shows *target appearance* with central calcified masses in the radiolucent lesion.
- **Radiopaque stage**—this shows multiple radiopacity continuous with the surrounding bone, but they were separated from adjacent teeth periodontal ligament space. In this stage *cotton wool appearance* (lobular or lump shaped and soft radiopaque characters like that of cotton wool) is seen.
- **Active hypercementosis**—hypercementosis of tooth in the affected area is seen. There is an also wide periodontal ligament space.

Differential Diagnosis

- **Paget's disease**—no radiolucent capsule and increased serum alkaline phosphatase levels. Paget's disease affects entire mandible while florid osseous dysplasia occur above the inferior alveolar canal.
- **Chronic sclerosing osteomyelitis**—signs of infection is present in osteomyelitis.
- **Osteopetrosis**—profuse thickening of the skull base or calvarium and diffuse bony radiopacities. It will cause enlargement of bone, which is not a feature of florid osseous dysplasia.

Diagnosis

- **Clinical diagnosis**—painless expansion in four quadrant of jaw will give clue to diagnosis.
- **Radiological diagnosis**—radiopaque, mixed lesion in all four quadrants with active hypercementosis will diagnose this condition.

Management

- **Effective oral hygiene**—if teeth present effective oral hygiene should be maintained since with this disease, patients exhibit poor healing and osteomyelitis may develop after tooth loss.
- **Recontouring**—patients with more severe form of the disease has superficial lesions which are located near the crest of alveolar ridge, these may require recontouring to accommodate the denture or to prevent ulceration.

Cemento-ossifying Fibroma

This is the terminology used for *cementifying fibroma* and *ossifying fibroma*. Cemento-ossifying fibroma is a benign fibro-osseous neoplasm. The description of the ossifying fibroma of the jaws was first given by Montgomery in 1927. It is a benign neoplasm that is osteogenic (non-odontogenic), well defined and rarely encapsulated, consisting of fibrous tissue with variable amounts of mineralized material similar to bone and/ or cementum. Chromosomal abnormalities have been identified in the ossifying fibroma; however, the molecular mechanisms that causes the development of this tumor remain unknown.

Nomenclature

The previous nomenclature of ossifying fibroma or cementifying fibroma was based upon the histopathology which displayed predominantly bone like or cementum like tissue, characterized by osteoblasts and cementoblasts respectively. This terminology was first used by Hamner, but is of little practical value because of the range of the behavior of these lesions with identical histopathological appearance. Furthermore many of them contained both bone and cementum like elements. Therefore these lesions

appear to represent points on a spectrum of histological appearances extending from bone to cementum. This indicates that they probably originated from the same progenitor cell, which Waldron suggested is to be found within the periodontium.

Taking account of the wide range of histological manifestation, the WHO in 1992 revised its nomenclature to refer to the separate lesions of the cementifying fibroma and ossifying fibroma as a single entity termed 'cemento-ossifying fibroma'.

Clinical Features

- *Age and sex distribution*—it predominantly occurs in females in third or fourth decades of life.
- *Bones affected*—it can arise from any part of the facial skeleton and skull with over 70% of cases arising in the head and neck region. Cemento-ossifying fibromas ostensibly do not occur outside the craniofacial complex. Although it has been principally found in the jaws, it has also been reported in the orbitofrontal bone, nasopharynx, paranasal sinuses and skull base. Most lesions arise in the tooth bearing areas of the mandible and maxilla (**Fig. 34-20**). It mostly occurs in molar and premolar mandibular area above the inferior alveolar canal. In the maxilla, it occurs most often in the canine fossa and zygomatic arch area.
- *Symptoms*—there is occasional facial asymmetry is seen in some of the cases. When in maxilla, symptoms may include nasal stuffiness and epiphora on the affected side.
- *Signs*—there may be associated exophthalmos, with visual disturbances, depending on the extent of compression of its orbital content by the tumor.
- *Giant ossifying fibroma*—large lesions increasing in size to over 80 mm in their greatest diameter have been termed 'giant ossifying fibroma' (**Fig. 34-21**).
- *Teeth*—the lesion is slow growing and in some cases, there is displacement of teeth.
- *Cortex*—bony cortex and covering mucosa remain intact.



Fig. 34-20: Cemento-ossifying fibroma involving maxillary lesion (Courtesy Dr Bhaskar Patle).



Fig. 34-21: Giant ossifying fibroma showing lesion over 80 mm in diameter.

- *Progress*—the lesion may be slow growing initially, with a rapid increase in size in a relatively short time.
- *Maxillary sinus*—if sinus is affected it may fill the sinus completely and expands the sinus wall.

Radiographic Features

- *Periphery*—the borders of cemento-ossifying fibroma lesions usually are well defined. A thin radiolucent line, representing a fibrous capsule, may separate it from surrounding bone. Sometimes, the bone next to the lesion develops a sclerotic border.
- *Internal structure*—it is a mixed radiolucent/ radiopaque density with a pattern that depends on the amount and form of the manufactured calcified material. In the early stages solitary cyst-like osteolytic lesions without periosteal reaction are seen. In some instances, the internal structure may appear almost totally radiolucent with just a tint of calcified material. In the type that mainly contains abnormal bone, the pattern may be similar to that seen in fibrous dysplasia, or a wispy (similar to stretched tufts of cotton) or flocculent pattern (similar to large, heavy snowflakes) may be seen (**Fig. 34-22**). Lesions that produce more cementum like material may contain solid, amorphous radiopacities (cementicles) similar to those seen in cemental dysplasia.
- *Effect on surrounding structure*—cemento-ossifying fibroma can be distinguished from the previously mentioned bone dysplasia by its tumor like behavior. This is reflected in growth of the lesion, which tends to be concentric within medullary part of bone with outward expansion approximately equal in all directions. These neoplasms have propensity for osseous cortical expansion and encroachment on contiguous structures. A significant point is that the outer cortical plate, although displaced and thinned remains intact. In the mandible, bowing or erosion of the inferior cortex is seen. It results in displacement of the inferior alveolar canal. The cemento-ossifying fibroma lesion can grow into and occupy the entire maxillary sinus, expanding its walls outward; however, a bony partition always



Fig. 34-22: Cemento-ossifying fibroma presenting a flocculent pattern.

exists between the internal aspect of the remaining sinus and the tumor. In the orbit they may displace the globe. Expanding lesion can cause displacement of the teeth. The lamina dura of involved teeth usually is missing and resorption of teeth may occur.

Diagnosis

- *Clinical diagnosis*—not so specific. Facial asymmetry is seen.
- *Radiological diagnosis*—mixed radiopaque radiolucent lesion seen with sclerotic border. Displacement of teeth can also be seen
- *Laboratory diagnosis*—large number of fibroblasts, with flat elongated nuclei is present within the network of interlacing collagen fibers. Chinese letter shaped islands of bone or calcification is also seen.

Differential Diagnosis

Early radiolucent stage

- *Post extraction socket and residual cyst*—history of extraction and history of surgery
- *Primordial cyst*—it is always associated with a missing permanent tooth which is not a case with cementifying/ossifying/cemento-ossifying fibroma.
- *Ameloblastoma*—it occurs in posterior most part of the mandible and is accompanied by paresthesia of the lip. It often shows a multilocular appearance.
- *Periapical cemental dysplasia*—it occurs at the apices of vital teeth. However, they affect an older age group than cementifying/ossifying/cemento-ossifying fibroma and have a predilection for the lower incisor region whereas cementifying/ossifying/cemento-ossifying fibroma often occurs in premolar-molar region.

- *Adenomatoid odontogenic tumor*—usually associated with impacted maxillary canines in a younger patient and borders are regular.

Mixed radiopaque radiolucent lesion

- *Calcifying epithelial odontogenic tumor*—it occurs in posterior body and ramus of mandible.
- *Osteoid osteoma*—it is more common in males under 30 years of age and is not frequently seen in jaws, whereas, cemento-ossifying fibromas are tumors predominantly of jaws and are common in females in their third or fourth decade of life. It causes pain that predominantly occurs in night time, which is a distinguishing feature as cemento-ossifying fibroma is mostly an asymptomatic lesion. Radiographically, it appears as a well-circumscribed radiolucent area with sclerotic halo in initial lesions. In more mature lesions, the central radiolucency may have a radiopaque foci representing abnormal bone.
- *Osteoblastoma*—it is rare lesion in the head and neck. Histologically, it exhibits bone to stroma ratio of one to one, with frequent fibroblastic atypia and occasional mitotic figures, features which help in differentiation from cemento-ossifying fibroma when radiographic features are confusing.
- *Osteosarcoma*—it should be differentiated on the basis of malignant characteristics of the tumor such as aggressive growth, destruction of cortical bone, invasion into surrounding soft tissues and along the periodontal ligament, whereas, cemento-ossifying fibromas has a benign behavior.
- *Benign cementoblastoma*—it occurs in second and third decades of life in posterior mandibular region. It is characteristically attached to the part of the root with frequent root resorption. Cemento-ossifying fibroma is not attached to the root although it may resorb the roots. Radiographically it is an opaque lesion with a radiolucent halo. Histological study reveals agglomerates of material similar to cementum and connective tissue stroma. Many of the cemental trabeculae are surrounded by cementoblasts. Clinically, it may sometimes manifest as a pain and swelling unlike cemento-ossifying fibroma, which is usually not associated with pain.
- *Juvenile ossifying fibroma*—Contrary to cemento-ossifying fibroma, which occurs mostly in third and fourth decades of life, juvenile ossifying fibroma appears in patients under 15 years of age. Cemento-ossifying fibroma most commonly affects mandible predominantly premolar-molar region, whereas, juvenile ossifying fibroma affects predominantly maxilla. It has a rapid growth that frequently erodes the surrounding bones. Though the radiographic appearance of this entity has similarities to cemento-ossifying fibromas, it shows differences histopathologically. Histological picture is characterized

by a cellular vascular stroma with variable amount of giant cells, scarce collagen and small ossicles surrounded by an osteoid halo. They do not have fibrous capsule.

- *Metastatic osteoblastic carcinoma and chondrosarcoma*—ill-defined borders with periosteal bone formation. It is rapidly growing, as compared to cementifying/ossifying/cemento-ossifying fibroma which has well defined margins and separated from normal adjacent bone by a radiolucent fibrous capsule.
- *Periapical cemental dysplasia*—as described in differential diagnosis of periapical cemental dysplasia.
- *Paget's disease*—cotton wool appearance and enlargement of affected bone.
- *Fibrous dysplasia*—homogeneous radiopaque area with an internal architecture that is evenly granular and obliterates normal bone marrow space. Borders are ill defined and gradually blend into surrounding normal bone. Root resorption is rare. It is more commonly found in mandible, with an equal sex distribution.

Mature stage

- *Periapical spherical type of hypercementosis*—it is attached to a part of root and is separated from the periapical bone by a radiolucent periodontal ligament space which surrounds the entire root.
- *Condensing osteitis*—it can be ruled out because it occurs at the periapex of a non-vital tooth. It does not have a radiolucent rim which is seen in fibro-osseous lesions.
- *Periapical idiopathic osteosclerosis*—it occurs in the periapical region of vital teeth. Cementifying/ossifying/cemento-ossifying fibroma however is smoothly contoured and almost round and ovoid, whereas periapical idiopathic osteosclerosis is usually quite irregular in shape and also there is absence of a radiolucent rim.
- *Complex odontoma*—density is not uniform and also it seldom occur periapically.

Management

- *Enucleation*—small, clinical encapsulated lesions are treated by conservatively enucleation.
- *Resection*—it is recommended if there is involvement of inferior border, extension into maxillary sinus occurs.

Peripheral Ossifying Fibroma

It presents as a tumor like growth of the oral soft tissues and is often associated with sharp teeth, rough restoration, and ill-fitting dentures.

Clinical Features

- Even though cemento-ossifying fibroma is considered as an intraosseous lesion, affects the gum and soft tissue have also been described.

- An excrescent, ulcerated, painful, friable lesion that bleeds when touched could be observed.
- Clinically, they usually present as reddish brown, firm, pedunculated or sessile masses at the site of trauma (Fig. 34-23).
- The overlying mucosa may be ulcerated and this may draw the patient's attention to the lesion. If the patient is dentate, the adjacent teeth may be mobile.



Fig. 34-23: Peripheral ossifying fibroma (Courtesy Dr Soni).

Radiographic Features

- In edentulous patients, there may appear to be some superficial erosion of the underlying bone on radiographs.
- Intra-oral films taken with low penetration may show varying amounts of calcification within the lesion.

Diagnosis

- *Clinical diagnosis*—not so specific.
- *Radiological diagnosis*—erosion of underlying bone below the lesion with calcification will give clue to diagnosis.

Other Lesions of Bone

Osteoporosis

There is reduction in the inorganic constituent of bone. There is abnormal persistence of calcified cartilage. Spongy portion of affected bone ultimately becomes a solid block of calcified cartilage leaving inadequate space for hemopoiesis. It is characterized by low bone mass and micro-architectural bone fragility. It is usually rarefaction of bone resulting from deficiency of bone matrix rather than deficit mineral.

Types

- *Primary* (associated with aging).
- *Secondary* (reduction of histologically normal bone that has been accelerated by abnormal or iatrogenic circumstance such as corticosteroid or heparin therapy or condition such as malnutrition and scurvy).

Mechanism of Bone Loss

- *Imbalance between bone resorption and bone formation*—it is caused by imbalance between bone resorption and bone formation with an exaggeration of resorption, reduction in bone formation or combination of both.
- *Postmenopausal and senile*—it occurs due to overall decrease in anabolic hormones (estrogen) in postmenopausal women. There may be lag in formation of bone and since bone resorption is continued, it results in osteoporosis. After the age of 60 years, generalized atrophy of bone occur (senile osteoporosis). Senile osteoporosis is caused by decrease in calcium absorption, vitamin D absorption and metabolism with age, which in turn decreases the anabolic hormones, muscular proteins and flow of blood to bone. By virtue of this condition, bone resorption occurs. Another cause of bone resorption in adults is that, in elderly person altered hormonal function lead to formation of small thrombi which plug small vessels in bone and cause loss of bone vitality and hence resorption.
- *Cushing's syndrome*—the symptoms are caused by an increased output of glucocorticosteroids, especially cortisol. The excess cortisol acts to produce osteoporosis by two ways:
 - It contributes to the degradation of proteins and severely limits the formation of bone matrix by reducing the amount that each osteoblast synthesizes.
 - It promotes the formation of osteoclasts from the osteogenic undifferentiated cells and thus enhances bone resorption.
- *Drug-induced osteoporosis*—prolonged administration of cortisol and cortisone show Cushing's like syndrome and cause osteoporosis in the same way as that in Cushing's syndrome. Contraceptive drug can also cause osteoporosis; as after administration of contraceptive drug, there is increase in level of cortisol. Combination of these two compounds leads to production of abnormal megakaryocytes, which in turn leads to formation of abnormal sticky platelets. These sticky platelets fuse to form thrombi which occlude small vessels in the tissue due to which the bone dies and get resorbed.
- *Malnutrition state*—sufficient protein must be absorbed from intestine to supply constant need for matrix formation. Deficiency causes osteoporosis and may result from protein poor diet or from GIT disturbances

such as colitis, regional arteritis. Vitamin C deficiency may weaken sinusoidal vessel walls in medullary bone which tend to dilate and rupture, resulting in pooling of blood and hypoxia, which leads to loss of vitality and removal of bone by physiologic osteoclast.

- *Stress*—there is interaction between the progesterone compounds and increased level of cortisol by stress, leading to formation of multiple small thrombi. The thrombi occlude small vessels of osseous tissue, which results in death and resorption of bone, leading to osteoporosis.
- *Thyrotoxic osteoporosis*—it is usually seen in children with hyperthyroidism. Thyroxin mediates the action of cortisol on bone and an excess of thyroxin results in a more efficient utilization of steroids. Thus there is increased resorption.
- *Osteoporosis and oral bone loss*—a decrease in bone mass from unbalanced system in which there is decreased bone formation or increased bone resorption, is designated as skeletal osteopenia. The designation of osteoporosis being given when the changes in bones are accompanied by pain, deformity or fracture. It is also defined as too little calcified bone and as a condition when the concentration of bone (mineral) lies more than two standard deviation below the corresponding values for the age, when matched with normal value.

Clinical Features

- *Age*—either present at birth (congenital) or developed in early life (infantile).
- *Fracture*—bones are fragile and susceptible to fracture. These fractures typically affect the forearm (Colle's fracture), spine (vertebral fracture) and hip (femur fracture).
- *Congenita form* inherited as an autosomal recessive disorder is invariably fatal in early life due to massive hemorrhage, anemia and rampant bone infections occurring due to progressive loss of bone marrow and their cellular products.
- *Percussion*—percussion over the affected vertebrae is painful.
- *Symptoms*—osteoporotic patient may notice gradual loss of height due to shortening of trunk. In advanced cases, clinical onset is often characterized by attack of severe pain which is aggravated by movements and occurs after trauma.
- *Osteomyelitis*—it may occur due to relatively avascular and there may be bone pain.

Oral Manifestations

- *Osteomyelitis*—marked predilection for development of osteomyelitis.

- *Jaws*—fracture of jaws during tooth extraction.
- *Enamel hypoplasia*—enamel hypoplasia. Microscopic dentinal defect.
- *Teeth*—arrested root development, retardation of tooth eruption. Delayed eruption and early loss of teeth missing teeth and malformed teeth. Teeth are poorly calcified and are prone to caries.

Radiographic Features

- *Appearance*—typical wedge appearance of affected vertebrae, on a lateral radiograph.
- *Internal structure*—reduction in number of trabeculae is least evident in the alveolar process. Persistent trabeculae tend to occur along planes of bone stress. Trabeculae may be arranged in a radial manner, with wide spaces in between.
- *Cortex*—reduced density and thinning of cortical boundaries, such as inferior mandibular cortex (**Figs 34-24 and 34-25**).
- *Lamina dura*—the lamina dura surrounding the teeth may appear thinner than normal
- Anatomic shadows such as the nasal fossa and maxillary sinus are less distinct.

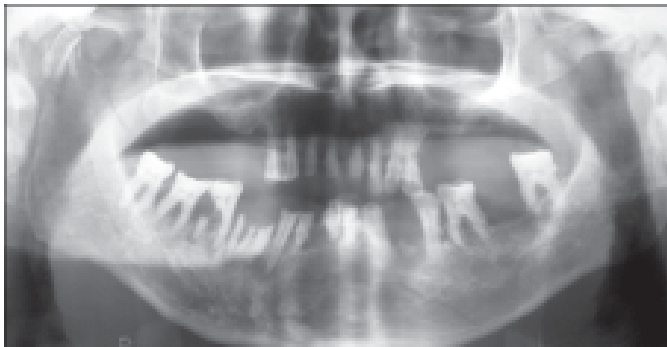


Fig. 34-24: Osteoporosis presenting as decrease in density of bone (Courtesy Dr Parate).

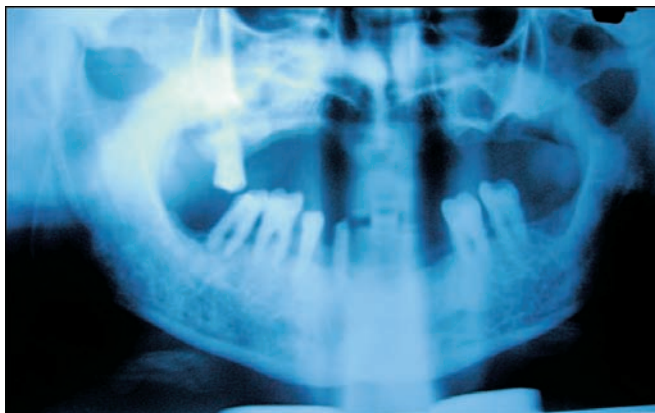


Fig. 34-25: Senile osteoporosis showing reduce density of the bone.

Diagnosis

- *Clinical diagnosis*—osteomyelitis of the jaw with fragile bone and fracture of bone will give clue to the diagnosis.
- *Radiological diagnosis*—wedge shaped appearance of the vertebrae with reduce density of bone in the jaw.
- *Laboratory diagnosis*—there is anemia, hepatomegaly, decrease RBC count and increase serum phosphatase levels. Biopsy shows lack of physiologic bone resorption. Osteoblasts are prominent but osteoclasts are seldom found. Trabeculae are in disorder arrangement.

Differential Diagnosis

- *Infantile cortical hyperostosis*—positive history of highly specific soft tissue and bony abnormalities are found. Sites of involvement limited to few bones. New bone is subperiosteal and continuous to inferior border.

Management

- *Life style changes*—good nutrition and regular exercise helps to prevent osteoporosis. Person should be asked to stop smoking and alcohol.
- *Calcium supplement*—they are widely used as an adjunct to other treatment in the prevention and treatment of osteoporosis. Recommended dose is 1000 mg/day.
- *Anabolic steroids* such as stanozolol, vitamin D and vitamin D active metabolites are also used.
- *Bisphosphonates*—new drug for systemic osteophonates are under evaluation and this includes bisphosphonates. These are synthetics analogue of pyrophosphate that absorb on to bone surface and become incorporated into the bone matrix. When the bone contains bisphosphonates is ingested by resorbing osteoclasts, the drug released within cell in high concentration and thus exerts a toxic effect, leading to cell death and inhibits the bone resorption.
- *Hormone replacement therapy*—hormone replacement therapy with estrogen is the treatment of choice of prevention of osteoporosis. Progesterone should be added to estrogen in women with intact uterus, to reduce the risk of endometrial carcinoma.
- *Alendronate* 110 mg daily with calcium supplement is also effective. Etidronate 400 mg daily for 2 weeks followed by 13 weeks calcium supplement is also useful.
- *Calcitonin injection*—injection of *calcitonin* to reduce bone resorption can be given. But it is rarely used due to side effects as hot flushes and nausea.
- *Grafting procedures* to rebuild the residual ridge and implants to stabilize dentures are being used in advanced cases of bone loss.
- *Prevention of bone loss*—oral bone loss can be prevented by plaque control, removal of local etiologic factors and use of antibiotics in case of early onset periodontitis.

Infantile Cortical Hyperostosis

It is also called as 'Caffey's disease', 'Caffey-Silverman syndrome'. It was first described in detail in 1945. It is characterized by unusual cortical thickening of certain bones and it occurs in two forms i.e. autosomal dominant form and sporadic form.

Etiology

- *Embryonic osteodysgenesis*—it may be an embryonic osteodysgenesis consequent to local defect in blood supply to the area.
- *Inherited defect*—Inherited defects of arterioles supplying the affected part results in hypoxia producing focal necrosis of overlying soft tissue and periosteal proliferation.
- *Allergy* as the basis of disease, edema and inflammation producing periosteal elevation and subsequent deposition of calcium.
- *Hereditary* being an autosomal dominant trait.

Clinical Features

- *Age and sex*—it is equal in males and females, with average onset at 9 weeks of age, with most of the cases arising before 6 months.
- *Sites*—bones commonly affected are mandible, clavicle, scapula, frontal bone and ulna. There is bilateral involvement in every case. Of all these bone clavicle and mandible are the most common sites.
- *Symptoms*—infants will develop fever and become hyper-irritable.
- *Signs*—soft tissue swellings have a sudden onset, especially in the facial area and early in disease, they may be warm and tender. Swelling is devoid of clinical signs of inflammation on the overlying skin. Swelling disappears slowly, but may suddenly recur at the same place or new site may be involved.
- *Others feature*—other features are pseudo-paralysis, pleurisy and dysphasia.

Oral Manifestations

- *Sites*—mandible is one of the commonest bones affected in infantile cortical hyperostosis.
- *Symptoms*—patient may have malocclusion and enamel hypoplasia may be present.
- *Signs*—residual asymmetric deformity of mandible, usually in the angle and ramus area.

Radiographic Features

- *Incidence*—only after swelling have subsided and ceased to be tender, many bone changes occur.

- *Cortex*—thickening of inferior cortex caused by new bone formation deep to the periosteum takes place.
- *Mandible*—overall enlargement of body of mandible with homogeneously increased density throughout.
- *Laminated appearance*—occasionally, the new bone may be laid down in layers giving the inferior cortex a laminated appearance.
- *Surface*—the surface of new bone may be smooth in some cases, while irregular in some cases.
- *Lamina dura*—the lamina dura and cortices of tooth follicle are normal.

Diagnosis

- *Clinical diagnosis*—infant with fever, soft tissue swelling, with swelling in the jaw will give clue to the diagnosis.
- *Radiological diagnosis*—thickening of cortex with laminated appearance.
- *Laboratory diagnosis*—anemia, leukocytosis, elevated erythrocyte sedimentation rate, monocytosis and increased serum alkaline phosphatase levels.

Differential Diagnosis

- *Callus formation*—unilateral or at least asymmetrical, when seen bilaterally.
- *Osteoma*—rarely discovered in a younger age.
- *Cherubism*—multilocular expansile radiolucency with no cortical thickening.
- *Osteoporosis*—in infantile cortical hyperostosis, abnormal bone production is subperiosteal while in osteoporosis, it is cancellous.
- *Osteomyelitis*—osteomyelitis of jaw may be associated with laying down of much subperiosteal new bone. But infantile cortical hyperostosis occurs within 6 months of life, as compared to osteomyelitis which occurs very unusually below 6 months of life. In osteomyelitis, whole of the mandible is not involved.
- *Periostitis of jaw*—it is apparently unilaterally involved and is localized. It is possible to see underlying cause of the condition. Laminated structure in periostitis is visible only at the inferior margins as compared to infantile cortical hyperostosis which is visible all over the bony surface of jaw.
- *Fibrous dysplasia*—Caffey's disease represent the changes added to be the bone, while fibrous dysplasia is an abnormality within the bone.
- *Hypovitaminosis A*—it produce the condition called as cortical hyperostosis. But it usually affects metacarpal bone and occurs over the age of 1 year, while infantile cortical hyperostosis occurs below 6 months and mandible is commonly affected.

Management

- There is no specific treatment for this disease.

Osteopetrosis

It is also called as 'Marble bone disease', 'Albers-Schonberg disease', 'osteosclerosis fragilis generalisata'.

It is a rare disorders characterized by an increase in density of bones, which becomes hard and brittle. This is a result of endosteal bone production with a lack of concomitant bone resorption and remodeling. The reason for this is that there is lack of normal osteoclast formation.

Types

- *Infantile osteopetrosis or malignant osteopetrosis*—it is a malignant childhood condition with a severe form showing severe skeletal and neurological signs. Those affected rarely live beyond the age of 2 years. It is autosomal recessive.
- *Intermediate osteopetrosis*—this is less severe form of osteopetrosis with fracture occurring at the end of first decade. It is autosomal recessive.
- *Transient osteopetrosis*—it shows radiographic evidence of diffuse sclerosis and it resolves without therapy. It is autosomal recessive.
- *Adult's osteopetrosis or benign osteopetrosis*—it is an autosomal dominant form. The adult form is characterized by predominantly skeletal signs with long having little or no defect.

Clinical Features

- *Age*—it can occur in infant and some cases are diagnosed later in life.
- *Skeletal features*—almost every one has a varying degree of poor skeletal development, if the disease appears early in life. Subsequent skeletal deformities occur later due to bending of the long bones and improper healing after pathological fractures.
- *Symptoms*—as a result of continuous bone deposition and lack of bone resorption, the foramina of cranial nerves are constricted, hence there is loss of hearing, disturbed vision, which diminish progressively.
- *Facial palsy*—facial nerve palsy is also seen.
- *Signs*—due to displacement of hematopoietic bone marrow, anemia ensues and the hematopoietic function is assumed by the liver, spleen and the lymph nodes resulting in hyperplasia of lymphoid tissues and hepatosplenomegaly. There is also frontal bossing, obliteration of maxillary sinus and possible hydrocephalus and mental retardation.
- *Prognosis*—prognosis of infantile osteopetrosis is very poor as compared to adult's osteopetrosis.

Oral Manifestations

- *Sites*—it is most common in mandible and occasionally, in maxilla.
- *Osteomyelitis*—osteomyelitis of jaws associated with osteopetrosis probably follows the obliteration and fibrosis of marrow is caused by reduced osseous circulation.
- *Paranasal sinus*—paranasal sinus may become readily obliterated.
- *Teeth*—there may be enamel hypoplasia, defective dentin, disturbed tooth development, small pulp and tendency towards caries.

Radiographic Features

- *Radiodensity*—there is diffuse homogeneous sclerotic appearance of all bones, within distinction between the cortex and the marrow. This type of appearance is called as marble bone appearance (Fig. 34-26).



Fig. 34-26: Homogeneous sclerotic appearance seen in marble bone disease (Courtesy Dr Ganju).

- *Skull changes*—the changes in the skull are often striking. The base of skull becomes grossly thickened and of great radiographic density. Loss of normal skull marking and structures. The dipoles are effaced, the head of patient resembles bladder of lard. There is gross thickening and increased opacity of the cranial base, with narrowing of foramina.
- *Fracture*—due to increased calcification there is increased fragility, so fractures are common.
- *Bones*—bones are shorter than normal, heavy, thick and deformed. The ends of long bone spay out.

- *Cortex*—the cortices are thickened and differentiation between them and adjacent bone is greatly lessened.
- *Internal structure*—less marked changes may produced are the increase in overall density but trabeculation may be apparent. The trabeculae may get thickened and the marrow spaces are correspondingly small.
- *Lamina dura*—the lamina dura is almost lost in the general density. Some time density is such that the roots are nearly invisible on dental radiographs.
- *Teeth*—the roots of the teeth may be obscured, partially or totally, depending upon the bone density.
- *Cortex*—the medullary cavity is replaced by bone and cortex is thickened.

Diagnosis

- *Clinical diagnosis*—hearing deformity, fracture of bone with skeletal deformity will give clue to the clinical diagnosis.
- *Radiological diagnosis*—marble bone appearance is seen which is typical of this disease.
- *Laboratory diagnosis*—red blood cell count below 1,000,000 cells per cu mm. Elevated serum acid phosphate levels have been reported in patients with benign dominant osteopetrosis. Biopsy shows prominent osteoblasts, but osteoclasts are seldom found.

Differential Diagnosis

- *Polyostotic fibrous dysplasia*—fibrous dysplasia usually involves a part of bone, rather than the complete bone. Hence, involved bone generally exhibits asymmetric enlargement.
- *Paget's disease*—it can be differentiated from osteopetrosis by its classic bones. It most often involves the skull, pelvis, vertebrae, femur, maxilla and mandible. Also, serum chemistry measurements show markedly elevated alkaline phosphate levels.
- *Sclerotic cemental masses*—have a stronger predilection for black women over 30 years of age. The most common salient feature of this disease is that only jaws are affected, radiograph of the jaws reveal radiopaque masses frequently rimmed by radiolucent borders.
- *Infantile cortical hyperostosis*—skeletal changes are subperiosteal and not endosteal. Mandible is more commonly involved.

Management

- *Bone marrow transplantation*—this is the treatment of choice in osteopetrosis patient. But to find matched donor is a difficult process.
- *Interferon gamma 1b*—if interferon gamma 1-b is given in combination with calcitrol will reduce the bone mass.

- *Other modalities*—corticosteroid, parathormone, macrophage colony stimulating factor and erythropoietin can also be given.
- *Supportive treatment*—this is given to combat infection, osteomyelitis. Antibiotics which are given in infection are fluoroquinolones and lincomycin. Hyperbaric oxygen therapy can also be given.
- *Precaution*—avoid major surgery in patients with osteopetrosis. Performing dental extraction osteopetrosis patient has a risk of osteomyelitis and jaw fracture.

Osteogenesis Imperfecta

It is also called as 'brittle bone', 'Lobstein disease'. It is a serious disease of unknown etiology. It represents a hereditary autosomal dominant trait.

Pathogenesis

There impairment of collagen maturation. Collagen is main part of bone, dentin, sclera, ligament and skin. So collagen impairment will result in change in this structure.

Types

- *Congenital or Vrolik's type*—it is present at birth.
- *Tarda or Lobstein's type*—it is recognized later in life. It is also called as osteopsathyrosis.

Clinical Features

- *Age*—many infants with this disease are stillborn or die shortly after birth.
- *Bone deformity*—patient is having bone deformity and hyper extensibility of the joint. There is also bowing of the bone (**Figs 34.27A and B**).
- *Fracture of bone*—extreme fragility and porosity of bones with an attendant proneness of fracture. Fracture heals readily but new bone is of similar imperfection. It is common for fracture to occur while the infant is walking or crawling. Hyperplastic callus formation, which may mimic osteosarcoma, take place.
- *Blue sclera*—there is occurrence of pale blue sclera which is thin; pigmented choroids show and produce the blue color (**Fig. 34-28**).
- *Signs*—there is deafness due to osteosclerosis; laxity of ligament and peculiar shape of the skull. There is also abnormal electrical reaction of muscle. Increased tendency for capillary bleeding.

Oral Manifestations

- *Dentinogenesis imperfecta*—most of the times, osteogenesis imperfecta is associated with dentinogenesis imperfecta.



A



B

Figs 34-27A and B: Patient of osteogenesis imperfecta showing bone deformity (Courtesy Dr Lambade).

- *Enamel hypoplasia*—sometimes, there is also hypoplasia of teeth. Sometimes there is class I malocclusion and greater incidence of impacted 1st and 2nd molars.
- *Deciduous teeth*—deciduous teeth are poorly calcified and semi-translucent or waxy. Appearance of teeth is faint dirty pink, half normal size, with globular crowns and relatively short roots in proportion to other dimension.
- *Malocclusion*—there is increased prevalence of malocclusion which is caused by maxillary hypoplasia.



Fig. 34-28: Blue sclera seen in patient of osteogenesis imperfecta.

Clinical Types

- *Neonatal lethal type*—it is characterized by multiple fractures in infants and the child seldom survives.
- *Severe non-lethal type*—disease is not evident until late childhood and patient shows fracture of bone with minimum trauma. Although the fractured bone heals up rapidly, considerable skeletal deformity and dwarfed stature often develop.
- *Moderate and deforming type*—it is associated with dentinogenesis imperfecta and blue sclera.
- *Mild and non-deformity type*—the patients are clinically normal, but they have increased tendency for bone fracture, due to trauma.

Radiological Features

- *Wormian bones*—patient may show wormian bone (bones in skull sutures) (Fig. 34-29).



Fig. 34-29: Wormian bone seen in osteogenesis imperfecta (Courtesy Dr Lambade).

- **Bone deformity**—there is skeletal deformities manifested by bowing of bone and fracture of bone (**Fig. 34-30**).
- **Internal structure**—the bone is osteoporotic, there is less density and trabeculae are fewer in number.
- **Appearance**—the chin is sharply pointed, as a result of softening of bone, leading to flattening of sides of the mandible.

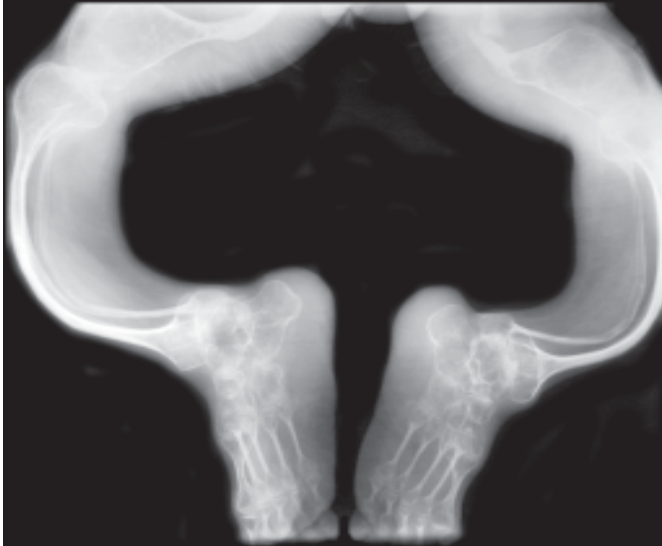


Fig. 34-30: Bowing of bone seen in radiograph of patient with osteogenesis imperfecta (Courtesy Dr Lambade).

Diagnosis

- **Clinical diagnosis**—severe bone deformity with blue sclera are diagnostic features of osteogenesis imperfecta.
- **Radiological diagnosis**—multiple wormian bones with osteopenia will diagnose this condition.
- **Laboratory diagnosis**—bone is composed of immature spongy bone. Osteoblastic activity retarded and there is failure of fetal collagen to be transformed into mature collagen.

Management

- **Management of fracture**—management of fracture should be done in patient.
- **Management of teeth**—teeth management should be same as that is carried out in dentinogenesis imperfecta.

Pierre Robin Syndrome

It is also called as '*Robin anomalad*'. It results arrested development.

Clinical Features

- **Triad**—it is triad of cleft palate, mandibular micrognathia and glossoptosis (causes airway obstruction).

- **Bird facies**—arrested development and ensuing hypoplasia of mandible ultimately produce the characteristic bird facies (**Fig. 34-31**). Mandibular hypoplasia yields a retrognathic appearance.



Fig. 34-31: Mandibular retrognathism resulting in characteristic bird face appearance.

- **Tongue**—the retruded mandible results in posterior displacement of the tongue.
- **Cleft palate formation**—abnormal descent of tongue occurs between the palatal shelves resulting in cleft palate (**Fig. 34-32**).



Fig. 34-32: Cleft palate formation seen in patient with Pierre Robin syndrome.

- **Respiration**—there is respiratory difficulty due to failure of support of tongue musculature because of micrognathia, allowing the tongue to fall down and backwards, partially obstructing the epiglottis. Difficulty in respiration is noted from birth and it can cause asphyxiation.

- *Others*—there may be congenital heart defects, other skeletal abnormalities and ocular lesions. Mental retardation is also a common finding.

Diagnosis

- *Clinical diagnosis*—triad of cleft palate, micrognathia and glossoptosis will diagnose this condition.

Management

- *Breathing support*—breathing support and feeding assistance are necessary during infancy.
- *Surgical*—surgical closure and orthodontic treatment are indicated.

Marfan's Syndrome

It is also called as 'Marfan-Achard syndrome', 'arachnodactyly'. It is hereditary disease transmitted as autosomal dominant trait. It is basically a disease of connective tissue related to defective organization of collagen which is abnormally soluble.

Clinical Features

- *Spider finger*—excessive length of the tubular bones resulting in disproportionately long, thin extremities, the finger and toes are long, thin and tapering so that the name 'spider finger' has been applied.
- *Shape*—the shape of face and skull is characteristically long and narrow.
- *Joint*—hyperextensibility of joint with habitual dislocations, kyphosis or scoliosis and flatfoot.
- *Bilateral ectopia lentis*—it is caused by weakening or rupture of the suspensory ligaments.
- *Cardiovascular complications*—cardiovascular complications like aortic aneurysm and aortic regurgitation, valvular defects and enlargement of the heart are common.

Oral Manifestations

- *Palate*—high arched palatal vault is very prevalent.
- *Maxilla and mandible*—bifid uvula, malocclusion and multiple odontogenic cysts of maxilla and mandible.
- *Temporomandibular joint*—there may be temporomandibular dysarthrosis.

Diagnosis

- *Clinical diagnosis*—spider finger, with high arched palate, hyperextensibility of joints.

Management

There is no specific treatment for this syndrome and prognosis is good.

Down's Syndrome

It is also called as 'Trisomy 21 syndrome' and 'mongolism'. It is associated with subnormal mentality in which an extremely wide variety of anomalies and functional disorders may occur. It results from excessive chromosomal material involving all or a portion of chromosome 21. It is caused by advanced maternal age, uterine and placental abnormalities and chromosomal aberration.

Type

- *Typical type*—trisomy-21 with 47 chromosomes (95 percent of cases).
- *Translocation type*—46 chromosomes.
- *Chromosomal mosaicism*.

Clinical Features

- *Skull*—flat face, large anterior fontanelle, open sutures. There is also brachycephalic skull shaped with frontal prominence and occipital flattening (**Figs 34-33A and B**).
- *Palpebral fissures*—they are almond shaped with superior-lateral or Mongolian obliquity.
- *Eyes*—small slanting eyes with epicanthal folds. Eyes are widely spaced. There is also ocular hypertelorism.
- *Nose*—there is flattened nasal bridge (**Fig. 34-33**).
- *Others*—sexual underdevelopment, cardiac abnormalities and hypermobility of the joints.



Figs 34-33A and B: Brachycephalic skull shaped with widely shaped eyes. Nose has flattened bridge.

Oral Manifestations

- **Tongue**—macroglossia with protrusion of tongue (**Fig. 34-34**), fissured tongue or pebbly tongue from enlargement of papilla.
- **Others**—open mouth, frequent prognathism and high arched palate.
- **Maxillary sinus**—there is hypoplasia or aplasia of maxillary sinus.
- **Teeth**—teeth are malformed, enamel hypoplasia and microdontia.
- **Malocclusion**—it attributable to small maxillary arch relative to the mandibular arch.
- **Periodontal disease**—universal severe destructive periodontal disease that do not appear to be local in origin.



Fig. 34-34: Malocclusion with macroglossia of tongue (Courtesy Dr Parate).

Radiological Features

- **Periodontal destruction**—periodontal destruction of bone can be observed on the radiograph (**Fig. 34-35**).

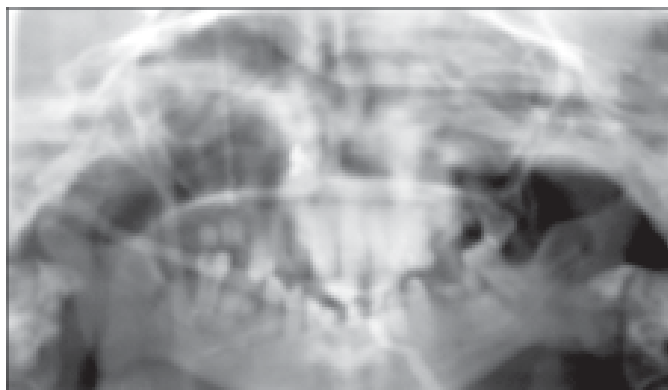


Fig. 34-35: Periodontal destruction of bone seen in the OPG (Courtesy Dr Parate).

Diagnosis

- **Clinical diagnosis**—wide open eyes with brachycephalic shaped with malocclusion will give clue to the diagnosis.
- **Radiological diagnosis**—it is not specific.

Management

There is no specific treatment for this syndrome and many patient died during first year of life.

Achondroplasia

It is also called as '*chondrodystrophia fetalis*'. It is a disturbance of endochondral bone formation, which results in a characteristic form of dwarfism. It is a hereditary condition, which is transmitted as an autosomal dominant trait.

Clinical Features

- **Appearance**—patient is quite short, usually less than 14 meters in height and thickened muscular extremities, brachycephalic skull and bowed legs.
- **Hands**—hands are usually small and fingers are stubby.
- **Skull**—the base of the skull is small and constricted as a result of retarded growth of the cartilaginous portions. The calvarium is large and bulges frontally and laterally.
- **Nose**—there is also depression of the bridge of nose.
- **Lumbar lordosis**—lumbar lordosis with prominent buttocks and protruding abdomen is often present.
- **Joints**—joints exhibit limitation of motion.
- **Others**—arms do not hang freely at the sides and the elbows can not be straightened.

Oral Manifestations

- **Prognathism**—maxilla is often retruded because of restriction of growth at the base of skull which may produce relative mandibular prognathism.
- **Teeth**—congenital missing teeth with disturbances in shape of teeth may also occur.

Radiographic Features

- **Shorter bone and bone clubbing**—long bones are shorter than normal and there is thickening or mild clubbing at the ends.
- **Epiphysis**—epiphysis may close, either early or late.
- **Narrow foramen magnum**—bones at the base of the skull fuse prematurely, producing shortening as well as a narrow foramen magnum.

Diagnosis

- **Clinical diagnosis**—short bone, depression of nose, malocclusion of teeth and prognathism will give clue to diagnosis.

- *Radiological diagnosis*—narrow foramina magnum and short bone
- *Laboratory diagnosis*—there is retardation or even aplasia of zone of provisional calcification of endochondral growth. Cartilage columns lack orderly arrangement, fail to calcify properly and are not resorbed and replaced by bone in the usual fashion.

Management

There is no specific treatment for this disease.

Skeletal Fluorosis (Fluoride Toxicity)

Fluoride toxicity can be manifested in two forms i.e. acute and chronic. It can be caused by mutation and other genetic disorders.

Clinical Features

- *Symptoms*—nausea, vomiting and epigastric distress. Excess salivation, diarrhea and mucus discharge with headache and sweating. There is pain in joints of hand, feet and spine.
- *Signs*—barely detectable pulse, hypotension, cardiac arrhythmias and disturbance in electrolyte balance. There is also stiffness of gait, limitation of movement occurs.
- *Severe cases*—in most severe cases, there is almost crippling of movement and extreme pain because the spine and joints become rigid and virtually, immobilization of the individual occurs.
- *Complication*—respiratory and metabolic acidosis and coma.

Radiographic Features

- *Bone density*—density of bone is increased.
- *Thickening of bone*—bones of extremities show thickening and osteophytes.
- *Calcification*—calcification occurs in tendons and ligaments.

Diagnosis

- *Clinical diagnosis*—excess salivation, headache, sweating and joint pain will give clue to diagnosis.
- *Radiological diagnosis*—calcification and thickening of bone.

Management

- *Elimination of fluoride from body*—all attempts should be to eliminate the toxic dose of fluoride from the body.
- *Maintenance of vital signs*—support all the vital signs.

- *Emetics*—administer emetics if the patient possesses a gag reflex i.e. if he is not vomiting.
- *Glucose*—glucose, to reverse hyperkalemia and calcium gluconate to maintain sodium and calcium levels must be given.
- *Sodium bicarbonate*—sodium bicarbonate or Ringer lactate must also be given to increase salivary flow, decrease acidosis, to increase pH and to increase excretion of fluoride.

Generalized Cortical Hyperostosis

It is also called as 'Von Buchem disease' and it represents an excessive deposition of endosteal bone throughout the skeleton in a pattern suggestive of a hereditary condition, with an autosomal recessive characteristic.

Clinical Features

- *Appearance*—facial appearance of this patient may be swollen, particularly with widening of the angles of mandible and at the bridge of nose.
- *Symptoms*—in some patients, there is loss of visual activity and loss of facial sensation; some degree of facial paralysis and deafness occurs and all this is due to cranial nerve involvement through closure of foramina.
- *Alveolar process*—intraorally, there is sometimes overgrowth of alveolar process.

Radiological Features

- *Increase density*—a skeletal radiograph reveals increased density of many bones of body.
- *Skull sclerosis*—the skull exhibits diffuse sclerosis which may be present in the jaws.

Diagnosis

- *Clinical diagnosis*—overgrowth of alveolar process, facial paralysis and deafness.
- *Radiological diagnosis*—skull sclerosis, and increases density of bone
- *Laboratory diagnosis*—the bone is normal dense bone, but without evidence of remodeling.

Management

There is no treatment although patient may live his normal life.

Massive Osteolysis

It is also called as 'vanishing bone', 'disappearing bone', 'phantom bone', 'progressive osteolysis' or 'Gorham syndrome'. It is characterized by spontaneous, progressive resorption of bone with ultimate total disappearance of bone.

Pathogenesis

In this condition, there is progressive destruction of bone. The bone is replaced by fibro-vascular tissue. It occurs due to proliferative vascular and connective tissue response to unknown mechanism.

Clinical Features

- *Age and sex distribution*—it is most common in older children, young and middle age adults. There is no sex predilection.
- *Sites*—the most commonly affected bones are the clavicle, scapula, humerus, ribs, ilium, ischium and sacrum.
- *Symptoms and progress*—the disease may or may not be painful, begins suddenly and advance rapidly, until the involved bone is replaced by a thin layer of fibrous tissue surrounding a cavity.

Oral Manifestations

- *Site*—intraorally mandible is more commonly affected as compared to maxilla.
- *Symptoms*—the patients may present with pain or facial asymmetry. In some cases there is history of gradual progressive recession of the lower jaw.
- *Pathological fracture*—there is pathologic fracture of bone following even minor trauma.
- *Signs*—there may be complete destruction of mandible (**Fig. 34-36**). Deviation of mandible can also occur in the affected side. Ulceration of the lesion is usually absent.



Fig. 34-36: Destruction of mandible seen with no ulceration in buccal mucosa in mandibular posterior region (Courtesy Dr Chandrasekhar Kadam).

Radiological Features

- *Radiodensity*—it has got osteolytic pattern radiologically.
- *Appearance*—radiolucency is present in the mandible showing complete destruction. Margins of the lesion are indistinct (**Fig. 34-37**).



Fig. 34-37: Vanishing bone disease showing destruction of bone on left side of the mandible (Courtesy Dr Chandrasekhar Kadam).

- *Lamina dura*—there is absence of lamina dura and thinning of cortical plates.

Diagnosis

- *Clinical diagnosis*—pain, facial asymmetry and pathological fracture without any apparent cause may suspect massive osteolysis
- *Radiological diagnosis*—it will show complete destruction of bone in the jaws
- *Laboratory diagnosis*—biopsy shows vascular proliferation with osteoclastic reaction.

Management

- *Surgical*—it can be managed by intra-periosteal or extraperiosteal resection followed by autogenous grafts.
- *Radiation therapy*—some authors suggest that the role of moderate dose of radiation therapy. It is one of most successful treatment for the massive osteolysis.
- *Zeledronic acid*—it has got anti-angiogenic properties which inhibits osteoclasts can be useful in resorptive process of vanishing bone disease.
- *Nutritional treatment*—vitamin D-50000 units/week, calcium glycerophosphate 6 gm/day and fluoride.
- *Other treatment*—various other treatment modalities like chemotherapy, calcitonin, mithramycin disphosphonates also has been tried but with limited success.

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Introduction

AIDS is a devastating fatal disease which is in epidemic form throughout the world. It is an incurable viral STD (sexually transmitted disease) which is caused by human immunodeficiency virus (HIV). It stands for:-

- *A*—Acquired, i.e. contagious not inherited.
- *I*—Immune, i.e. power to resist disease.
- *D*—Deficiency.
- *S*—Syndrome, i.e. number of signs and complaints indicative of a particular disease.

The case of AIDS was detected in *June 1981* when five young homosexual men came with the suffering from rare lung infection due to microorganism called *Pneumocystis carinii*. In India, the first description of AIDS came in Chennai where six women out of 125 who were screened were HIV positive in high risk group of prostitutes.

HIV attacks the immune system of the body. Due to that an individual is not able to protect himself from potentially harmful organism.

AIDS appears to be endemic in central and equatorial Africa and it may be old disease of Africa that has gone unrecognized. HIV infection has also become the primary emphasis of effort at controlling STDs. Moreover, the knowledge gain about sexual and other behavior associated with transmission of HIV, as well as strategies that have been effective in modifying those behaviors, is transferable to other sexually transmittable and blood-borne infections. These above factors have revolutionized standard approaches to the control of these infections.

Oral and perioral lesions are common in patients with human immunodeficiency virus, which are often the presenting feature and may have deterioration of general health and a poor prognosis.

Definition

WHO has given the following definition of AIDS.

- One or more opportunistic infections listed in clinical features that are at least moderately indicative of underlying cellular immune deficiency.
- Absence of all known underlying causes of cellular immune deficiency (other than HIV infection) and absence of all other causes of reduced resistance reported to be associated with at least one of those opportunistic diseases.

Prevalence

It is more common in African country and Western countries particularly in the United States. The largest population of AIDS occurs in homosexuals, intravenous drug users, and heterosexuals with sexual contact with AIDS patient. It can also occur in patients who received transfusion of blood or blood pigments donated by the person with risk factors. About 92% of victims are males, 6.5% females with 1% children. It is common in the age group of 25 to 49 years.

Transmission

- *Sexual transmission*—it is in 90% of cases. It depends upon number of sexual partners, receptive anal intercourse and presence of other STDs. All these are in high risk group. Prostitution is a major heterosexual factor associated with AIDS. Homosexuals have more risk of transmitting HIV than the heterosexual males.
- *Use of contaminated blood products*—intravenous drug users, HIV-contaminated blood transfusion, blood clotting concentrate and organ transplantation.
- *Perinatal transmission*—it occurs in 13% among children born to HIV seropositive mother.
- *Other nosocomal routes*—transmission from patient to patient due to reuse of contaminated and shared needles.

- *Professional hazards*—the risk of transmission from HIV-infected patient to health care workers is more than health care workers to patient.
- *Saliva transmission*—HIV has been found to be present in saliva. Saliva reduces the ability of HIV to infect its target cells. So transmission through saliva is rare possibility. But, there are some rare example which documented that HIV is transferred from the saliva of the patient.

History of Nomenclature of Virus

- *T lymphocytes*—there is quantitative and qualitative deficiency of T₄ helper cells in AIDS patients. This lead to certain investigators to focus their efforts on determining if etiologic agent was a virus that manifested a particular tropism for T₄ helper lymphocytes.
- *HTLV-III virus*—Dr Robert C Gallo determined that type C retrovirus was tropic for T₄ lymphocytes in adult T-cell leukemia/lymphoma. He named the virus, *Human T cell leukemia/lymphoma virus (HTLV-I)*. So, it is considered to be etiological agent for AIDS. It causes lymphoproliferation in T cell leukemia, whereas AIDS is a disease of lymphodepletion. The answer came in the discovery of type D retrovirus of HTLV family that has been termed as HTLV-III.
- *LAV virus*—on the other hand, virus called lymphadenopathy associated virus (LAV) was isolated from the AIDS patient in Europe.
- *Cytopathic human T lymphocytotropic virus*—HTLV III and LAV are closely related members of same class of virus. Finally it is proved that HTLV and LAV are cytopathic human T-lymphocytotropic viruses that manifested selective infectivity for the helper/inducer subset of T cells that as phenotypically designated reactivity with monoclonal antibody T₄ or Leu₃.
- *HIV*—in order to avoid different nomenclatures, retrovirus responsible for the AIDS are named 'Human immunodeficiency virus' which belong to family of retroviruses.

Characteristic of the HIV Virus

HIV virus is lymphotropic virus and its primary target is T₄ cell. HIV is a spherically enveloped virus, about 90-120 nm in size which is much smaller than bacteria. The nucleocapsid has an outer icosahedral shell and inner cone-shaped core, enclosing the ribonucleoproteins.

The genome is diploid, composed of two identical single stranded, positive sense RNA copies. Inside the envelops, there is a protein core, which contains enzymes reverse

transcriptase, integrase, protease etc. all essential for viral replication and maturation.

When the virus infects a cell, the viral RNA is transcribed by the enzymes, first into single-stranded DNA and then to double-stranded DNA (provirus) which is integrated into the host cell chromosomes. The virus is extremely sensitive to heat, thus boiling and autoclaving are very effective measures of inactivating the virus.

Mechanism of Action

- *Normal mechanism*—pathogenic viruses → identified by macrophage → it activates T lymphocytes → it gets differentiated into effector cell like T helper cell (T₄) and T suppresser cell (T₈) → T₄ cells secrete various lymphokines which induce lymphocyte to be differentiated into plasma cell → it secretes specific antibodies against viral antigen → it destroys the virus.
- *Mechanism in AIDS*
 - *Antigenic stimulation*—when the virus enters the blood stream, it evokes antigenic stimulation which activates CD4, T helper lymphocytes and macrophages.
 - *Secretion of TNF and interleukin 6*—lymphocytes and macrophages then secrete growth factors, tumor necrosis factor (TNF) and interleukin-6. This results in increase in CD4 cells thereby increasing number of cells vulnerable for HIV virus.
 - *Adherence of virus to CD4 cells*—HIV virus then adheres to CD4 surface via interaction between viral gp120 surface glycoprotein and CD4 cell surface membrane receptors. This surface interaction occurs with the help of transmembrane protein (chemokine receptor-5 or CCR-5).
 - *Replication of HIV virus*—after this, HIV virus will replicate in CD4 cell which also proliferates by antigenic stimulus.
 - *Incubation period*—once the viral genes are integrated into cells of own DNA, they can apparently remain dormant for an indefinite period of time, without causing any effects. This is called as 'incubation period'.
 - *Fate of CD4*—initially CD4 cells reduce the number of HIV virus by cellular and humoral attacks. But as continuous replication of virus occurs in lymph nodes, it will destroy the lymph nodes. This will lead into unchecked replication of virus which will lead to decrease in CD4 count of the human being. When the number of CD4 count is severely depleted below 200/mm³, the immune system collapses and variety of infections occur. At this stage, the patient is said to have AIDS.
- *Window period*—it is time between entry of virus into the body and the blood test becoming positive. This period

is about 1 to 3 month and person is infectious during that period.

Six Groups at Risk of Developing AIDS

- Homosexuals or bisexuals—71.4%
- Intravenous drug users—18.4%
- Hemophilia
- Recipient of multiple blood transfusion
- Infant born of parents belonging to first 3 high risk groups
- Heterosexual contacts of high risk group.

Classification

1993 Revised Classification System for HIV Infection

The Centers for Disease Control and Prevention (CDC) has revised the classification system for HIV infection to emphasize the clinical importance of the CD4+, T-lymphocyte count in the categorization of HIV-related clinical conditions. Consistent with the 1993 revised classification system, CDC has also expanded the *AIDS surveillance case definition* to include all HIV-infected persons who have less than 200 CD4+, T-lymphocytes/ μ L, or a CD4+, T-lymphocyte percentage of total lymphocytes of less than 14. This expansion includes the addition of three clinical conditions *pulmonary tuberculosis, recurrent pneumonia, and invasive cervical cancer*.

Measures of CD4+, T-lymphocytes are used to guide clinical and therapeutic management of HIV-infected persons. The revised CDC classification system for HIV-infected adolescents and adults categorizes persons on the basis of clinical conditions associated with HIV infection and CD4+, T-lymphocyte counts. The system is based on three ranges of CD4+, T-lymphocyte counts and three clinical categories.

CD ₄ + T cell categories	A Asymptomatic, acute HIV and PGL	B Symptomatic, not A or C conditions	C AIDS indicator condition
Category 1 > 500/ μ L (>28%)	A ₁	B ₁	C ₁
Category 2 : 200 to 499/ μ L (14-28%)	A ₂	B ₂	C ₂
Category 3 < 200/ μ L (<14%)	A ₃	B ₃	C ₃

Category A

It consists of one or more of the conditions listed below in an adolescent or adult (greater than or equal to 13 years)

with documented HIV infection. Conditions listed in Categories B and C must not have occurred.

- *Asymptomatic HIV infection*
- *Persistent generalized lymphadenopathy*
- *Acute (primary) HIV infection with accompanying illness or history of acute HIV infection*

Category B

It consists of symptomatic conditions in an HIV-infected adolescent or adult that are not included among conditions listed in clinical Category C and that meet at least one of the following criteria:

- The conditions are attributed to HIV infection or are indicative of a defect in cell-mediated immunity.
- The conditions are considered by physicians to have a clinical course or to require management that is complicated by HIV infection.

Examples of conditions in clinical category B include, but are not limited to:

- *Bacillary angiomatosis*
- *Candidiasis, oropharyngeal (thrush)*
- *Candidiasis, vulvovaginal; persistent, frequent, or poorly responsive to therapy*
- *Cervical dysplasia (moderate or severe)/cervical carcinoma in situ*
- *Constitutional symptoms, such as fever (38.5°C) or diarrhea lasting greater than 1 month*
- *Oral hairy leukoplakia*
- *Herpes zoster (shingles), involving at least two distinct episodes or more than one dermatome*
- *Idiopathic thrombocytopenic purpura*
- *Listeriosis*
- *Pelvic inflammatory disease, particularly if complicated by tubo-ovarian abscess*
- *Peripheral neuropathy*

For classification purposes, Category B conditions take precedence over those in Category A. For example, someone previously treated for oral or persistent vaginal candidiasis (and who has not developed a Category C disease) but who is now asymptomatic should be classified in clinical Category B.

Category C

Conditions included in the 1993 AIDS surveillance case definition

- *Candidiasis of bronchi, trachea, or lungs*
- *Candidiasis, esophageal*
- *Cervical cancer, invasive **
- *Coccidioidomycosis, disseminated or extrapulmonary*
- *Cryptococcosis, extrapulmonary*
- *Cryptosporidiosis, chronic intestinal (greater than 1 month's duration)*

- Cytomegalovirus disease (other than liver, spleen, or nodes)
- Cytomegalovirus retinitis (with loss of vision)
- Encephalopathy, HIV-related
- Herpes simplex: chronic ulcer(s) (greater than 1 month's duration); or bronchitis, pneumonitis, or esophagitis
- Histoplasmosis, disseminated or extrapulmonary
- Isopsoriasis, chronic intestinal (greater than 1 month's duration)
- Kaposi's sarcoma
- Lymphoma, Burkitt's (or equivalent term)
- Lymphoma, immunoblastic (or equivalent term)
- Lymphoma, primary, of brain
- Mycobacterium avium complex or *M. kansasii*, disseminated or extrapulmonary
- Mycobacterium tuberculosis, any site (pulmonary * or extrapulmonary)
- Mycobacterium, other species or unidentified species, disseminated or extrapulmonary
- Pneumocystis carinii pneumonia
- Pneumonia, recurrent *
- Progressive multifocal leuko-encephalopathy
- Salmonella septicemia, recurrent
- Toxoplasmosis of brain
- Wasting syndrome due to HIV

*Added in the 1993 expansion of the AIDS surveillance case definition. (This classification is in adapted form of CDC).

Walter Reed Staging System

The Walter Reed staging system of HIV infection classifies patients on the basis of CD4 counts, skin-test responsiveness, lymphadenopathy, oral candidiasis and opportunistic infections. It has limitations with respect to predictive value.

WHO Clinical Staging of HIV/AIDS and Case Definition

The clinical staging and case definition of HIV for resource-constrained settings were developed by the WHO in 1990 and revised in 2007. Staging is based on clinical findings that guide the diagnosis, evaluation, and management of HIV/AIDS, and does not require a CD4 cell count.

Primary HIV Infection

- Asymptomatic
- Acute retroviral syndrome

Clinical Stage 1

- Asymptomatic.
- Persistent generalized lymphadenopathy.

Clinical Stage 2

- Moderate unexplained weight loss (<10% of presumed or measured body weight).
- Recurrent respiratory infections (respiratory tract infections, upper respiratory infections, sinusitis, tonsillitis, bronchitis, otitis media, pharyngitis).
- Herpes zoster.
- Minor mucocutaneous manifestations (angular cheilitis, recurrent oral ulcerations, seborrheic dermatitis, prurigo, papular pruritic eruptions, fungal fingernail infections).

Clinical Stage 3

- Unexplained severe weight loss (>10% of presumed or measured body weight).
- Unexplained chronic diarrhea for >1 month.
- Unexplained persistent fever for >1 month (> 37.6°C intermittent or constant).
- Persistent oral candidiasis (thrush).
- Oral hairy leukoplakia.
- Pulmonary tuberculosis within the last 2 years.
- Severe presumed bacterial infections (e.g. pneumonia, empyema, pyomyositis, bone or joint infection, meningitis, bacteremia).
- Acute necrotizing ulcerative stomatitis, gingivitis or periodontitis.
- Conditions for which confirmatory diagnostic testing is necessary.
- Unexplained anemia (hemoglobin <8 g/dL).
- Neutropenia (neutrophils <500 cells/ μ L).
- Chronic thrombocytopenia (platelets <50,000 cells/ μ L).

Clinical Stage 4

- HIV wasting syndrome, as defined by the CDC.
- *Pneumocystis jiroveci* (formerly *carinii*) pneumonia.
- Recurrent severe or radiological bacterial pneumonia.
- Chronic herpes simplex infection (oral or genital, or anorectal site) for >1 month.
- Esophageal candidiasis.
- Extrapulmonary tuberculosis.
- Kaposi's sarcoma.
- Central nervous system toxoplasmosis.
- HIV encephalopathy.

Conditions for which a confirmatory diagnostic testing is necessary:

- Cryptococcosis, extrapulmonary
- Disseminated non-tuberculosis *Mycobacterium* infection.
- Progressive multifocal leukoencephalopathy
- Candidiasis of the trachea, bronchi, or lungs
- Chronic cryptosporidiosis
- Chronic isosporiasis

- Visceral herpes simplex infection, cytomegalovirus infection (retinitis or organ other than liver, spleen, or lymph node)
- Any disseminated mycosis (e.g. histoplasmosis, coccidioidomycosis, penicilliosis)
- Recurrent nontyphoidal *Salmonella* bacteremia
- Lymphoma (cerebral or B-cell non-Hodgkin's)
- Invasive cervical carcinoma
- Visceral leishmaniasis

USPHS–CDC Classification

This classification is given by United State Public Health Service Center of Disease Control

- *Group I*—acute infection
- *Group II*—asymptomatic infection
- *Group III*—persistence generalized lymphadenopathy.
- *Group IV*—other disease
 - *Subgroup A*—constitutional diseases
 - *Subgroup B*—neurological diseases
 - *Subgroup C*—secondary infectious diseases
- *C₁*—specified secondary infectious diseases listed in CDC surveillance definition for AIDS.
- *C₂*—other specified secondary infectious stages:
 - *Subgroup D*—secondary cancer
 - *Subgroup E*—Other conditions

AIDS-related Complex

In some persons before the development of obvious AIDS, some clinical and laboratory findings are present. This is called AIDS-related complex. For clinical and research studies, persons exhibiting complex clinical problems and immunological or hematological abnormalities on the laboratory tests, have been classified as having AIDS-related complex (ARC). **ARC** requires any two or more symptoms and two or more abnormal laboratory findings. It must be present for at least 3 months.

- *Clinical finding*—it includes lymphadenopathy, weight loss of 15 lbs or 10% of body weight, fever of 38.5°C which is intermittent or continuous, diarrhea, fatigue, malaise, night sweats and oral candidiasis.
- *Laboratory finding*—there is decreased number of T helper cell, decreased ratio of T helper cells to T suppressor cells. There is also anemia, leucopenia, thrombocytopenia, lymphopenia, increased serum globulin level, decreased blastogenic response of lymphocytes to mitogen and increased level of circulating immune complex.
- *Cutaneous anergy*—cutaneous anergy to multiple skin test antigens. Anergy is impaired or inability to react to skin antigens.

Clinical Features

- *Acute symptoms*—they are more or less like infectious mononucleosis. Patient complaint of sore throat, fever, headache, myalgia, arthralgia, diarrhea, photophobia, maculopapular rash and peripheral neuropathy.
- *Protozoa and helminthes infection*—the most common opportunistic infection is by *Pneumocystis carinii* which causes pneumonia, CNS infection or other disseminated infections and toxoplasmosis, cryptosporidiosis (intestinal) causing diarrhea for over one month
- *Fungal infection*—*Candida* organism may cause esophageal candidiasis, bronchial or pulmonary candidiasis. Cryptococcosis causing CNS infection. Patients may suffer from disseminated histoplasmosis.
- *Bacterial infections*—*Mycobacterium avium* intracellulare causing infection disseminated beyond lung and lymph node. *Mycobacterium tuberculosis* causing tuberculosis.
- *Viral infections*—cytomegalovirus, causing infection in the internal organs other than liver, spleen and lymph nodes. Herpes simplex virus, causing chronic mucocutaneous infection with ulcers persisting more than one month.
- *AIDS dementia complex*—this is most common manifestation of AIDS. It is progressive encephalopathy.
- *Persistent generalized lymphadenopathy*—in silent phase patients may have persistent generalized lymphadenopathy. Most frequent sites involved in PGL are cervical, occipital and axillary lymph nodes.
- *Malignancy*—Kaposi's sarcoma is the most common vascular malignancy seen in AIDS patient. Kaposi's sarcoma usually occurs in homosexuals. Another malignancy which may occur is non-Hodgkin's lymphoma which most commonly affects CNS.

Oral Manifestations

Oral manifestations of HIV disease are common and include oral lesions and novel presentations of previously known opportunistic diseases. Careful history taking and detailed examination of the patient's oral cavity are important parts of the physical examination and diagnosis requires appropriate investigative techniques. Early recognition, diagnosis and treatment of HIV-associated oral lesions may reduce morbidity. The presence of these lesions may be an early diagnostic indicator of immunodeficiency and HIV infection, may change the classification of the stage of HIV infection and is a predictor of the progression of HIV disease. About 95% of AIDS patients have head and neck lesion and about 55% have important oral manifestation. They are depicted in **Table 35-1**.

Table 35-1: Oral disorders in HIV disease

Fungal*More common*

- Candidiasis

Less common

- Aspergillosis
- Histoplasmosis
- *Cryptococcus neoformans*
- Geotrichosis

Bacterial*More common*

- HIV gingivitis
- HIV periodontitis
- Necrotizing gingivitis

Less common

- *Mycobacterium avium intracellulare*
- *Klebsiella pneumoniae*
- *Enterobacterium cloacae*
- *E. coli*
- *Salmonella enteritidis*
- Sinusitis
- Exacerbation of apical periodontitis
- Submandibular cellulitis

Viral*More common*

- Herpes simplex
- Varicella zoster
- Epstein-Barr including hairy leukoplakia

Less common

- HPV virus
- CMV virus
- Pox virus

Neoplasm*More common*

- Kaposi's sarcoma

Less common

- Non-Hodgkin's lymphoma
- Squamous cell carcinoma

Lymphadenopathy**Neurologic disorders***Less common*

- Paresthesia
- Facial palsy
- Hyperesthesia
- Dysphagia

*Miscellaneous**Less common*

- Recurrent aphthous ulceration
- Progressive necrotizing ulceration
- Toxic epidermolysis
- Delayed wound healing
- Thrombocytopenia
- Xerostomia and sicca type syndrome
- HIV embryopathy
- Hyperpigmentation
- Granuloma annulare
- Exfoliative cheilitis
- Lichenoid and other drug reaction.

Common Oral Manifestations in AIDS

Candidiasis

Candidiasis is a common finding in people with HIV infection. Reports describe oral candidiasis during the acute stage of HIV infection, but it occurs most commonly with falling CD4+ T-cell count in middle and late stages of HIV infection. Several reports indicate that most persons with HIV infection carry a single strain of *Candida* during clinically apparent candidiasis and when candidiasis is inactive.

Clinical Features

- *Site*—patient with HIV usually has lesion of hard palate, tongue and soft palate.
- *Clinical pattern*—there are mainly four clinical pattern of candidiasis seen in AIDS patient. They are pseudomembranous, hyperplastic, erythematous and angular cheilitis.
- *Symptoms*—these lesions may be associated with a variety of symptoms, including a burning mouth, problems in eating spicy food and changes in taste. All three of these common forms may appear in one individual.
- *Pseudomembranous candidiasis (thrush)*—characteristic creamy white, removable plaques on the oral mucosa are caused by overgrowth of fungal hyphae mixed with desquamated epithelium and inflammatory cells. The mucosa may appear red when the plaque is removed. This type of candidiasis may involve any part of the mouth or pharynx.
- *Erythematous candidiasis*—Erythematous candidiasis appears as flat, red patches of varying size. It commonly occurs on the palate and the dorsal surface of the tongue. Erythematous candidiasis is frequently subtle in appearance and clinicians may easily overlook lesions, which may persist for several weeks if untreated.
- *Angular cheilitis*—it appears clinically as redness, ulceration and fissuring, either unilaterally or bilaterally at the corners of the mouth. It can appear alone or in conjunction with another form of candidiasis.
- *Hyperplastic candidiasis*—in this, candidal lesion has got firm appearance. It usually does not scrape on pressure. (Fig. 35-1).

Management

- *Topical clotrimazole*—it is the treatment of choice. Patient improves a lot after applying topical clotrimazole application.
- *Topical itraconazole*—itraconazole in as a oral rinse is also effective in controlling oral candidiasis in HIV patient.



Fig. 35-1: Hyperplastic candidiasis seen in patient with AIDS (Courtesy Dr Chole).

- **Systemic azoles**—it is given when patient is not receiving effective retroviral therapy and CD4+ count below 50% most commonly azoles which are used are fluconazole, ketoconazole, and itraconazole. All these drugs should be given carefully as they are associated with a lot of drug interaction.

Atypical Periodontal Disease

Periodontal disease is a fairly common problem in both asymptomatic and symptomatic HIV-infected patients.

Types

- **Linear gingival erythema**—also called *HIV-related gingivitis*. There is distinctive linear band of erythema (Fig. 35-2).
- **Necrotizing ulcerative gingivitis**—ulceration and necrosis of interdental papillae occurs.



Fig. 35-2: Linear gingival erythema (Courtesy Dr Soni).

- **Necrotizing ulcerative periodontitis**—also called *HIV-associated periodontitis* in which rapid loss of periodontal attachment is seen.

Clinical Features

- **Linear gingival erythema**—it often occurs in clean mouths where there is very little plaque or calculus to account for the gingivitis. In linear gingival erythema, the gingiva may be reddened and edematous. In some cases, alveolar gingiva and mucosa may demonstrate punctate diffuse erythema.
- **Necrotizing ulcerative gingivitis**—Patients sometimes complain of spontaneous bleeding. In acute-onset ulcerative gingivitis, ulcers occur at the tip of the interdental papilla and along the gingival margins (Fig. 35-3). It often elicits severe pain. The ulcers heal, leaving the gingival papillae with a characteristic *cratered* appearance.



Fig. 35-3: Ulcerative gingivitis showing ulceration at the margin of gingiva.

- **Necrotizing ulcerative periodontitis**—necrotizing ulcerative periodontitis may present as rapid loss of supporting bone and soft tissue (Fig. 35-4). Typically, these bone/tissue loss occur simultaneously with no formation of gingival pockets, sometimes involving only isolated areas of the mouth. Teeth may loosen and eventually fall out, but uninvolved sites can appear healthy.
- **Necrotizing stomatitis**—it may develop and areas of necrotic bone may appear along the gingival margin. The bone may eventually sequestrate. This resembles noma.
- **Difference between HIV-related periodontitis and non-HIV-related periodontitis**—the patient's history and clinical appearance make the diagnosis. It is sometimes difficult to distinguish this type of periodontal disease from non-HIV-related periodontal disease. However, the comp-

laints of severe pain, rapid onset and rapid destruction in an often extremely clean mouth are unusual for non-HIV-related periodontal disease.



Fig. 35-4: HIV-associated periodontitis showing extensive loss of periodontal support.

Management

- **Debridement**—removal of necrotic tissue should be carried out. After necrotic tissue is removed, irrigation with povidone-iodine solution is done.
- **Maintenance**—patient should be done periodic scaling and root planning. Oral hygiene should be maintained with a chlorhexidine mouth rinse once or twice daily.
- **Antimicrobial therapy**—in cases of NUP, metronidazole (250-mg four times daily), amoxicillin/clavulanate (Augmentin 250-mg three times daily), or Clindamycin (300-mg three times daily) should be given.

Herpes Simplex Infection

- **Forms**—it mainly appears as herpes labialis and recurrent intraoral herpes.
- **Herpes labialis**—herpes labialis occurs as characteristic lip lesion consisting of vesicles on an erythematous base that heals within 7 to 10 days.
- **Recurrent intraoral herpes**—these lesions are more widespread as compared to lesion in the non-HIV patients.
- **Treatment**—antiviral drugs and symptomatic treatment should be given. Systemic Acyclovir 30 mg/kg/day should be given. In acyclovir resistance patient, Foscarnet should be given.

Herpes Zoster

- **Occurrence**—it occurs more frequently in HIV-infected patients and carries poor prognosis.

- **Appearance**—the occurrence of unilateral vesicles that break and scab is characteristic of this infection (Fig. 35-5). This lesion persists for longer period of time than the normal course.
- **Oral features**—these conditions when present intraorally will lead to bone sequestration and loss of teeth. Pain is very severe nature.
- **Management**—acyclovir 15-30 mg/kg/day intravenously should be given for 8 hourly for 10 to 15 days.



Fig. 35-5: Intact vesicle seen in case of herpes zoster (Courtesy Dr Chole).

Hairy Leukoplakia

Oral hairy leukoplakia, which presents as a non-movable, corrugated or “hairy” white lesion on the lateral margins of the tongue occurs in all risk groups for HIV infections, although less commonly in children than in adults. It occurs in about 20% of persons with asymptomatic HIV infection and becomes more common as the CD4+ T-cell count falls below 200/mm³. Hairy leukoplakia is almost always a manifestation of HIV infection and clinicians should arrange evaluation of HIV disease.

Etiology

- **Epstein-Barr virus**—exact etiology is not known but Epstein-Barr virus (EBV) has been identified in these lesions. One hypothesis is that basal epithelial cells of lateral margin of tongue normally harbors EBV in majority of adult population, who are EBV seropositive and carrier of that disease. It is found primarily in homosexual males. Direct infection of Langerhan’s cell due to HIV-induced loss of factor essential for their integrity and function, permit reactivation of EBL with frequent epithelial hyperplasia.

Clinical Features

- **Site**—unique and significant lesion which primarily occurs unilaterally or bilaterally on the lateral border of tongue. It can also occur on dorsum of the tongue, buccal mucosa, floor of mouth, retromolar area and soft palate.
- **Appearance**—there is characteristic corrugated and white appearance. It does not rub off and may resemble the keratotic lesion.
- **Surface**—the surface is irregular and may have prominent folds (**Fig. 35-6**) or projections, sometimes markedly resembling hairs. Occasionally, however, some areas may be smooth and flat.
- **Spread**—they may also spread downward onto the ventral surface of the tongue, where they usually appear flat.
- **Pseudohairy leukoplakia**—Sometimes, white lesion satisfies many criteria for diagnosis of hairy leukoplakia, but if EBV is not present this is called 'pseudohairy leukoplakia'.



Fig. 35-6: Hairy leukoplakia present on the lateral border of tongue giving folded and hairy appearance.

Management

- **Antiviral drugs**—hairy leukoplakia usually is asymptomatic and does not require treatment. It disappeared in patients receiving high-dose acyclovir (2.5 to 3 mg per day for 2 to 3 weeks). Another antiviral drug which can be used is descyclovir.
- **Zidovudine**—HIV therapy with zidovudine affect EBV and significant regression of hairy leukoplakia occurs.
- **Topical treatment**—topical treatment with retinoid and podophyllin resins also gives temporary remission of hairy leukoplakia.
- **Others drugs**—other drugs which can use in hairy leukoplakia are phosphonoformate, topical clotrimazole (10 mg 5 times a day), nystatin (10000 units/gm 5 times a day) and ketoconazole (200 mg BD) and azidothymidine.

Kaposi's Sarcoma

It is also called '*angioreticulo-endothelioma*'. It is neoplasm of vascular endothelial origin. It is the most common tumor associated with AIDS and occurs in 1/3rd of AIDS patients.

Pathogenesis

- **Angiogenesis protein**—higher incidence of Kaposi's sarcoma is in homosexual men with AIDS as compared to heterosexuals with AIDS. It has been suggested that there is transmissible agent prevalence in homosexual population, which stimulates certain factors such as angiogenesis protein that may be critical in the pathogenesis of neoplasm. The patient with AIDS often shows clustered lesion in the oral cavity which suggests direct inoculation of mucosa with sexually transmitted agent.
- **Cytomegalovirus infection**—some theories suggest role of cytomegalovirus in the pathogenesis of Kaposi's sarcoma, but studies on prevalence of antibodies to cytomegalovirus in patient with classic and epidemic Kaposi's sarcoma have failed to demonstrate role of cytomegalovirus.
- **HHV₈ viruses**—nowadays, human herpes virus 8 (HHV₈) is thought to be in association with Kaposi's sarcoma in AIDS patient.

Types

- **Classic type**—it is rare neoplasm and occurs in older men. Usually, it appears as blue-black macule on the lower extremities. It is slow growing and rarely involves the lymph nodes and visceral organs.
- **African Kaposi's sarcoma**
 - **Cutaneous or aggressive variety**—it is considered an endemic disease and affects children. It appears as exophytic growth located in legs and arms. This form is locally aggressive and lymph node's involvement is rare.
 - **Nodular type**—it is similar to classic type.
 - **Florid variety**—it is characterized by rapidly progressive and widely disseminated lesions with visceral involvement.
 - **Lymphadenopathic variant**—the lymphadenopathic form occurs in children of 10 years age. The visceral and massive nodal involvement is common.
- **Kaposi's sarcoma with AIDS**—it is common in homosexuals but can occur in all risk groups. Male to female ratio is 20:1. Generally affects skin, oral and visceral organs. It may be the first symptom of AIDS.
- **Iatrogenic immunosuppression-associated Kaposi's sarcoma**—it is most common in recipient organ transplant. It occurs due to loss of cellular immunity.

Clinical Features

- **Site**—it occurs commonly in head and neck region. Tip of nose is peculiar and frequent location of it. It involves lymph nodes, soft tissue, extremities, gastrointestinal tract (GIT), lung, liver, pancreas, spleen and adrenal gland.
- **Age**—it can occur at any age but most common in 5th, 6th, 7th decade except in Africa where it occurs in children. It occurs most commonly in men but also has been observed in women.
- **Appearance**—it begins as multinucleated neoplastic process that manifests as multiple red or purple macules and in more advanced stage, a nodule occurring on the skin or mucosal surface.
- **Size**—it ranges from a few millimeters to a centimeter or more in diameter and is usually tender on palpation.

Oral Manifestations

- **Site**—it has tendency to involve the oral cavity, with hard palate as the most common site. But lesions may occur on any part of the oral mucosa including the gingiva, soft palate, buccal mucosa and in the oropharynx. It can involve either alone or in association with skin and disseminated lesions.
- **Appearance**—it can appear as a red, blue, or purplish lesion. It may be flat or raised, solitary or multiple. Occasionally, yellowish mucosa surrounds the lesion. The lesions may enlarge, ulcerate and become infected.
- **Symptoms**—patient may complain of pain and interference of lesion with eating and speaking
- **Signs**—lesions are tender and painful on palpation. After some period, the area may develop in plaque and nodule. In some cases, necrosis of tissue occurs.
- **Size**—it may vary in size from few millimeters to a centimeter or more in diameter.
- **Teeth mobility**—neoplasm can invade the bone causing mobility of the tooth.

Management

- **Oral prophylaxis**—it is important to perform thorough dental prophylaxis before initiating therapy for lesions involving the gingiva. Response to therapy is improved if all local plaque and calculus are removed.
- **Systemic chemotherapy**—systemic chemotherapy like vinblastine, vincristine, etoposide, alpha interferon, adriamycin, actinomycin D, and doxorubicin should be given.
- **Intralesional sclerosing agents**—intralesional injection of sodium tetradecyl sulfate has been effective for oral lesion.
- **Intralesional vinblastine**—it is useful for treating small lesions, particularly on the palate or gingiva. Several

studies have documented the effectiveness of one or two injections of 0.1 to 0.2 mg per ml solution of Vinblastine. Post-treatment pain is fairly common, but systemic effects are rare. The pain usually disappears several days after therapy.

- **Removal of lesion**—lesion may be removed by surgical excision, cryotherapy, LASER ablation, and electrocautery.
- **Radiation therapy**—it may be indicated for large, multiple lesions. A single dose of 800 cGy or an equivalent fractionated dose is frequently used and produces a good response. Side effects include xerostomia and mucositis, although both conditions usually improve with cessation of radiation therapy.
- **Antiretroviral therapy**—drugs such as indinavir, ritonavir, and saquinavir may cause significant regression in Kaposi's sarcoma.

Uncommon Oral Manifestation in AIDS

Human Papilloma Virus Lesions

- **Clinical features**—it is caused by human papilloma virus. It can cause *Verruca vulgaris* (common wart) and oral squamous papilloma. HPV lesions in the oral cavity may appear as solitary or multiple nodules. They may be sessile or pedunculated and appear as multiple, smooth-surfaced, raised masses resembling focal epithelial hyperplasia or as multiple, small papilliferous or cauliflower-like projections. It can be found on any mucosal surface and are contagious to both host and sex partner.
- **Management**—local excision of oral wart should be carried out. Recurrence is common. Other treatment modalities which are used are topical podophyllin, interferon, cryosurgery, laser ablation and electrocoagulation.

Cytomegalovirus Infection

- **Clinical features**—it has got predilection for salivary glands because many HIV-infected patients have xerostomia. It is a hypothesis that in such patient's salivary glands, cytomegalovirus infection produces inflammation causing reduced salivary production.
- **Management**—systemic ganciclovir or Foscarnet should be given.

Aphthous Ulcer

- **Clinical features**—aphthous ulcer are reflection of immune dysfunction. Report says that pituitary suppresses the

reduced host cortisone production, might account for this lesion. It is more commonly seen on tongue (Fig. 35-7). Major and herpetiform aphthous ulcer is more commonly seen as compared to minor ulceration.

- **Management**—intralesional injection of corticosteroid give successful results. Systemic steroid should be avoided to prevent further immunosuppression. Topical corticosteroid can also be given in this patient. In case of resistance cases of aphthous ulcer, thalidomide should be given. Thalidomide should be used for short-term as it may enhance the production of HIV.



Fig. 35-7: Aphthous ulcer seen on tongue in patient with AIDS (Courtesy Dr Amit Parate).

HIV-associated Salivary Gland Disease

- **Clinical features**—salivary gland disease associated with HIV infection can present as xerostomia with or without salivary gland enlargement. It usually involves parotid gland. The enlarged salivary glands are soft but not fluctuant. Patient may complain of discomfort due to the enlarged gland. HIV-infected patients may also experience dry mouth in association with taking certain medications that can hamper salivary secretion, such as antidepressants, anti-histamines and anti-anxiety drugs.
- **Diffuse infiltrative lymphocytosis syndrome (DILS)**—in this, there is CD8 lymphocytosis, lymphadenopathy and salivary gland enlargement.
- **Management**—removal of the enlarged parotid glands is rarely recommended. For individuals with xerostomia, the use of salivary stimulants such as sugarless gum or sugarless candies may provide relief. Candies that are acidic should be avoided as frequent use may lead to loss of tooth enamel. The use of salivary substitutes may also be helpful. For DILS, oral prednisolone and anti-retroviral therapy should be given.

Idiopathic Thrombocytopenic Purpura

- **Clinical features**—reports have described idiopathic thrombocytopenic purpura in HIV-infected patients. Oral lesions may be the first manifestation of this condition. Petechiae, ecchymosis and hematoma can occur anywhere on the oral mucosa. Spontaneous bleeding from the gingiva can occur and patients may report finding blood in their mouth on waking.
- **Management**—it best responds to anti-retroviral therapy.

Molluscum Contagiosum

- **Clinical features**—it is skin disease caused by pox virus. Lesions are waxy, dome-shaped papule with central crater. Facial skin also becomes commonly involved.
- **Management**—it is managed by curettage, cryosurgery and cautery. Recurrence is common. Resolution of lesion is also seen with anti-retroviral therapy.

Lymphoma

- **Clinical features**—most of the lymphoma present are non-Hodgkin's lymphoma. CNS involvement is seen with lymphoma. Oral lesion presents as soft tissue enlargement of the palate or gingiva. There is also loss of periodontal structure resulting in mobility of tooth.
- **Management**—it is managed by chemotherapy and radiation therapy.

Squamous Cell Carcinoma

- **Clinical features**—this can occur at younger age. HIV accelerated the development of oral squamous cell carcinoma.
- **Management**—it is managed by surgical removal, radiation therapy and chemotherapy.

Guidelines to Prevent Transmission to Dentist

- **Redesigning of instrument**—call for 'redesign' of instrument of surgical procedures that carry high risk of intra-operative injury to clinical staff.
- **Sterilization**—the understanding that all instruments used in dental procedures must either be sterilized before further use or be discarded.
- **Waste management**—elimination and protection from blood contaminated aerosols. Hazardous clinical waste should be disposed properly by the waste management.
- **Management of injuries**—protocol for management of needle stick and other intra-operative injuries aiming for defining the risk in given circumstances and administration of appropriate vaccine or other preventive measures.

- *Knowing HIV antibody status of patient*—there are several circumstances in which, it is important for dentist to know HIV antibody status for ensuring accurate differential diagnosis and appropriate treatment of the oral lesions those are associated with development of AIDS. This is also needed to assess the risk to dental health care worker after needle stick or other injury from contaminated once by contact with patient's blood and saliva.

Prevention

- *Educational counseling*—education counseling of general public should be done. It will help for increasing the awareness about AIDS.
- *Sexual restriction*—one should avoid sexual contact with suspect and in high-risk group. Multiple sex partners, intimate kissing and oral contact should also be avoided.
- *Disposable syringe and needles*—one should use disposable syringes and needles.
- *Blood transfusion*—blood donor should be properly screened.
- *Safety measure*—you should also educate health-care workers on safety measures.

Diagnostic Test for AIDS

ELISA (Enzyme-linked Immunosorbent Assay)

- *Mechanism*—it is a color reaction test in which prepared whole HIV virus particle which acts as antigen, will bind with antibodies to HIV virus in an infected human serum. A serum developed from the patient's blood sample, containing antibodies, is added to the ELISA plate, which is then washed clear of inactive antibodies that will not bind to antigens. A second layer of antibodies, called a conjugate, is added to detect the primary antibodies from the human serum. Excess antibodies are again washed clear of the plate, and finally, a substrate (chromogen) is added to make reactions occur.
- *Positive test*—if *positive*, the enzyme on the antibodies, once bound to the HIV antigens, will act on the substrate, changing its color (darker color means more serum has bound to the antigen).
- *Negative test*—if *negative*, no antibodies will bind to the HIV antigens, so no enzyme will be present to change the color of the substrate (no visible change will occur).
- *Advantage*—ELISA test is simple and convenient to perform; it is very sensitive for small amount of HIV antibodies and in initial screening tests.

- *Accuracy*—it is not 100% reliable because it is not completely specific only for HIV antibodies and reacts with other related viral antibodies and other non-specific chemicals giving false-positive result of about 5 to 25 per 1000 patients. In case of positive test, there is strong indication of past exposure and infection with virus but this does not show whether the patient is infectious because it does not show whether virus is still active or has been destroyed. If ELISA is positive, more accurate western blot method should be carried out.
- *Errors*—possible errors in the ELISA test may lead to a 'false positive' or a 'false negative' result. Reasons for 'false positives' include: women who have had multiple pregnancies (who may possess antibodies directed against human leukocyte antigens, confusing the test), or the presence of other diseases such as Lyme disease, lupus, or syphilis. "False negatives" (a test failure to register the presence of HIV), may occur during the early stages of HIV infection, before seroconversion (an antibody response to the virus) has occurred.

Western Blot Method

The Western blot test is a more specific version of the ELISA, which not only indicates whether a patient is HIV-positive or negative; it allows one to see which antibodies are directed against each viral protein. It is given as a follow-up to a positive ELISA test.

- *Mechanism*—in an HIV Western blotting, viral proteins from a blood sample are passed through a gel. Different proteins migrate through the gel at different speeds; typically, smaller proteins migrate through the gel faster than larger proteins. The separated proteins are then passed through an electric current so that they can transfer on a solid film strip in order of their speed. Human serum is added, and any existing HIV antibodies will bind to the HIV antigens. A chemical that reacts on contact with a protein-antibody-enzyme layer changes band color, just as in the ELISA test. Interpreting results is complicated, but the common "3-band rule" says that if three or more bands appear, HIV antibodies have been detected.
- *Positive test*—it is indicated when the treated i.e. HIV specific nitrocellulose strips are exposed to infected human serum and a goat antihuman antibody strip displays the characteristic band detected for each of three (env, pol and gag) group of viral protein.
- *Indeterminate test*—those that react with only one of these three groups of antigens are termed indeterminate.
- *Negative*—in this, it does not react with any viral protein.
- *Advantage*—it is more specific for HIV antibodies and is used to eliminate false-positive result.

Viral Load Testing

The viral load test measures the amount of HIV virus in the body. A viral load test should not be taken as a diagnostic test. Along with the CD4 cell count, the viral load test is used to give information about the progression of HIV infection, to predict its future course, and to guide recommendations for treatment.

- **Mechanism**—there are two types of viral load tests: PCR (polymerase chain reaction), and bDNA (branched DNA). The tests use different techniques to measure the same thing, which is the amount of HIV present in bloodstream. However, it is very important to use the same type of viral load test every time and not to switch between the two, since the values on test reports from the two different types are not comparable.
- **Reports**—the test report will show the number of HIV copies in one milliliter of blood. This indicates the current level of HIV, and the rate at which it is reproducing. The higher the count and reproductive rate, the faster the disease is likely to progress.
- **High viral load**—a high viral load can be anywhere between 5,000 and one million copies or more.
- **Low viral load**—a low viral load is usually between 200 and 500 copies. However, a low viral load only indicates slow progression, not an absence of HIV or cured HIV.
- **Significance of rate of change**—the rate of change in viral load between successive tests is also important. An increasing count indicates that a worsening infection, while a decreasing count indicates suppression of the HIV infection and improvement of the patient's health.
- **Errors**—there is a chance of a “false positive” result, indicating a falsely high viral load, especially when done by the very sensitive PCR method. There is also a chance of a “false negative” when the result is undetectable; an undetectable result does not mean the patient is cured of HIV.

Alternative Diagnostic Test

- **Indirect immunofluorescence and microfiltration enzyme assay system**—it is rapid and relatively inexpensive.
- **P24 antigen capture assays**—it is used for detection of HIV DNA that may be integrated into host DNA. HIV antibody can be detected by P24 antigen capture assay within 45 days after exposure.
- **Surrogate marker**—the absolute CD₄ + T cell lymphocyte count correlate best with progression of HIV-I related immune dysfunction. Other serum neoprotein beta-2-microglobulin HIV P₂₄ antigen interleukin-2 receptor IgA and impaired delayed type of sensitivity are also used.

Management of AIDS

- **Interferon**—it has got antiviral, antiproliferative and immunomodulator activity. The interferon is a glycoprotein produced by a number of different types of cells. Type I interferon (alpha and beta) are produced by leukocytes and fibroblasts. Type II interferon (gamma) is produced by lymphocytes and monocytes. Low doses of interferon enhance the antibody formation and lymphocyte blastogenesis. They also prolong cell cycle and cause inhibition of intracellular enzyme system (anti-neoplastic effect). The gamma interferons stimulate macrophage oxidative metabolism and have antimicrobial effect.
- **Thymic replacement therapy**—thymic epithelium plays an important role in transformation of blood-borne precursor cell into mature T cells. Thymic hormone or factor mediates this effect, since the immune system in AIDS is characterized by numerical and functional defects of T cell lymphocytes, it will correct the immune defect. Transplant of fetal thymus of cultural thymic epithelium and injection of thymic hormone have been successfully utilized in treatment of AIDS.
- **Lymphokines and cytokines**—lymphokines are materials produced by lymphocyte. Interleukin-I is macrophage product. In ‘*in vitro*’ system, interleukin-I enhances plaque forming cells responses and the generation of cytotoxic T cell alloantigen. In the presence of macrophage, interleukin-1 stimulates the production of interleukin-2, which stimulates and maintains the growth of T cell activated by antigens. Various studies have conformed that purified interleukin-2 (which stimulates and maintains growth of T cell activated by antigen), preparation *in vitro* system can normalize lymphocyte reaction in high percentage of individuals with unexplained lymphadenopathy and immunologic abnormalities, but the results are not significant in patients with AIDS.
- **Bone marrow transplantation**—syngeneic (identical twin) allogeneic bone marrow transplantation has been successful in reconstituting immune function in the patients with severe congenital immune defects. If this could be therapeutic in patient with AIDS that have appropriate marrow donor.
- **Monoclonal antibodies therapy**—In this, antibodies are directed against T cell differentiation antigens as a result of that number of circulating leukemic cells are decreased in patients with adult T cell active lymphoblastic leukemia.
- **Intravenous immunoglobulin therapy**—it reduces incidence of bacterial and viral infections. Infusion of hyper-immune gamma globulin enriched for neutralizing antibodies for LAV/HTLV III could be beneficial for

individuals with AIDS or ARC who have inadequate specific antibodies.

- *HAART (highly active antiretroviral therapy)*—this therapy altered the course of epidemic. Due to this therapy, survival rate of patients is increasing. Initial therapy consists of two nucleoside reverse transcriptase inhibitor and one or two protease inhibitor. Alternatively, two nucleoside reverse transcriptase inhibitor and non-nucleoside reverse transcriptase inhibitor can be given.
- *Nucleoside reverse transcriptase inhibitor*—it includes abacavir, didanosine, lamivudine, stavudine, zalcitabine and zidovudine. These drugs inhibit viral replication by competing with natural nucleoside when HIV virus synthesizes proviral DNA using its reverse transcriptase enzyme. As a result, faulty proviral DNA does not transcribe HIV RNA and replication is blocked.
- *Non-nucleoside reverse transcriptase inhibitor*—it includes didanosine, efavirenz, and nevirapine. These drugs act as non-competitive inhibitors of reverse transcriptase enzyme.
- *Protease inhibitor*—it includes amprenavir, indinavir, nelfinavir, ritonavir and saquinavir. It acts by blocking non-infective virus that immune system can destroy.

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Introduction

Hormones vary tremendously in chemical composition and in biologic activity. Various disorders of components of the endocrine system have generalized adverse effects on skeletal system due to altered metabolism.

Anatomy and Physiology

The endocrine system is specifically designed to integrate and control the human body's innumerable metabolic activities. Its functioning components are the endocrine glands. These units can function individually, in series, or in parallel, their activities being integrated closely. Communication with each other and with the tissues under their control is established by means of hormones, which they produce, store, and release as required and which are distributed throughout the body by means of the circulating blood.

In most instances, the agent stimulating or inhibiting their activity is the hormone produced by the corresponding target gland. The exceptions are the adrenal medulla, the posterior pituitary gland, and to a lesser extent, the pancreas. These glands are connected with the autonomic nervous system, secreting their hormones in response to electrical stimuli originating in higher centers in the brain and reaching them by way of the nerve fibers that link them to that system.

Pituitary Gland

- *Location*—it lies within the sella tursica at the base of brain and it is divided into three distinct lobes.
- *Lobes of pituitary gland*—the anterior lobe also called as adenohypophysis originates from epithelium of Rathke's pouch, the intermediated lobe from dorsal

portion of Rathke's pouch and posterior lobe or neurohypophysis develops from base of third ventricles.

- *Hormone secreted by anterior lobe*—hormone secreted by anterior lobe are growth hormone (GH), adrenocorticotrophic hormone (ACTH), thyroid stimulating hormone (TSH), follicle stimulating hormone (FSH), leutinizing hormone (LH) and prolactin.
- *Hormone secreted by intermediated lobe and posterior lobe*—hormone secreted by intermediated lobe is melanocytes stimulating hormone and by posterior lobe vasopressin and oxytocin.
- *Complex feedback interaction*—the secretory activities of the pituitary gland are modulated by hypothalamus through a series of complex feedback interaction.

Thyroid Gland

- *Location*—it is situated in midline of body in the neck, at the level of cricoid cartilage having two lateral lobes which are joined by isthmus. Third pyramidal lobe also extends from the isthmus.
- *Development*—embryologically the thyroid gland develops as a downgrowth from the portion of four pharyngeal pouches.
- *Function*—it regulates the basal metabolic rate, stimulates somatic and psychic growth and plays an important role in calcium metabolism.
- *Secretion from gland*—follicular cells lining the follicles of the gland secrete tri-iodothyronin and tetra-iodothyronin (thyroxin) which stimulates basal metabolic rate and somatic and psychic growth of the individuals. Para follicular cells lie in between the follicles and they secrete thyrocalcitonin which promotes deposition of calcium salts in skeletal and other tissue and tends to produce hypocalcemia.
- *Prohormone*—eighty-three percent of T_3 is produced by monodeiodination of T_4 in others tissue such as liver,

muscle and kidney. T_4 is probably not metabolically active until converted to T_3 and may be regarded as prohormone.

- **T_3 and T_4** — T_3 and T_4 circulate in plasma protein almost entirely bound to transport plasma proteins mainly thyroxin binding globulin. It is a minute fraction of unbound or free hormone, which diffuse into tissues and exerts its metabolic action. Production of T_3 and T_4 in the thyroid is stimulated by thyrotrophin (thyroid stimulating hormone—TSH), a glycoprotein released from thyrotrophic cells of the anterior pituitary in response to the hypothalamic tripeptide, thyrotrophic releasing hormone. There is negative feedback of thyroid hormones on the thyrotrophs such that in hyperthyroidism, when plasma concentration of T_3 and T_4 are raised, TSH secretion is suppressed and in hypothyroidism due to disease of the thyroid gland low T_3 and T_4 are associated with high TSH level.

Parathyroid Glands

- **Location**—the four parathyroid glands lie behind the lobes of the thyroid. They are not regulated by pituitary gland, but respond directly to changes in serum ionized calcium concentration.
- **Parathyroid hormone**—parathyroid hormone (PTH) is a single chain polypeptide of 84-amino acid which are synthesized by the chief cells and released in response to a fall in serum ionized calcium concentration.
- **Function**—PTH directly promotes reabsorption of calcium from renal tubules and bones. PTH also has indirect effect, mediated by increasing conversion of 1, 25 hydroxy cholecalciferol, which results in increase calcium absorption from the food and enhanced mobilization of calcium from bone. The initial effect of PTH on bone is to stimulate osteolysis, returning from bone to extracellular fluid.

Adrenal Gland

- **Location**—the adrenals are triangular-shaped structure that sits on the superior poles of the kidneys.
- **Secretion**—it produces and secretes a number of compounds that are essential for maintenance of life adaptation to stress.
- **Parts of gland**—each gland is divided into adrenal medulla and a cortex.
- **Adrenal medulla**—it arises from ectodermal tissues and function as a part of the sympathetic nervous system. It manufactures and secretes two catecholamine, i.e. epinephrine and norepinephrine.
 - **Epinephrine**—it supports blood pressure by increasing the heart rate. Epinephrine also increases oxygen consumption by the tissue and glucose release by the liver.

- **Norepinephrine**—norepinephrine increases peripheral resistance by its vasoconstrictor effect.
- **Metabolic effect**—epinephrine and norepinephrine also exert important metabolic effects by promoting lipolysis; increase blood sugar levels by stimulating glycogenolysis, elevating body temperature and increases basal metabolic rate. These compounds aid the body in adapting to stress which is important in the dental sitting because release of endogenous epinephrine during stressful dental procedure can produce significant changes in blood pressure and pulse rate.
- **The adrenal cortex**—it secretes three major classes of hormone: glucocorticoids or cortisols, which affects the inflammatory process and carbohydrates and protein metabolism, the mineralocorticoid, aldosterone, which affect water and electrolyte balance and sex hormone testosterone, estrogen and progesterone.

Diseases of Pituitary Gland

Hyperpituitarism

It results from hyperfunction of anterior lobe of pituitary gland, most significantly with increased production of growth hormone. The usual cause of this condition is a benign, functioning tumor of the eosinophilic cells in the anterior lobe of the pituitary gland. GH acts directly on some tissue but most of its biological effects are accounted by stimulation of secretion of insulin like growth factor I (IGF-I) and its binding proteins from the lower.

Types

- **Gigantism**—if the increase in level occurs before the epiphysis of the long bone are closed.
- **Acromegaly**—if the increase occurs later in life after epiphysis closure.

Clinical Features

Gigantism

- **Stature of individual**—generalized overgrowth of most tissue in childhood. Most of the soft tissue and bones respond to the excess hormone by enlarging. Excessive generalized skeletal growth can occur. Patient may often have of height to 7 to 8 feet. Patients achieve monstrous size because of tumors of the pituitary gland.
- **Symptoms**—later in life it may show genital underdevelopment and excessive perspiration and they complained of headache, lassitude, fatigue, muscle and joints pain and hot flashes.
- **Skull**—there is increase in size of calvarium which may lead to change in the hat size (**Fig. 36-1**). Pituitary tumors may also induce deficiency of other pituitary hormones

causing signs of hypogonadism including decreased libido and menstrual problems in women.

Acromegaly

- *Age and sex*—it is more common in males and occurs most frequently in 3rd decade.
- *Symptoms*—there is temporal headache, photophobia and reduction in vision.
- *Facial features*—bone overgrowth and thickening of the soft tissue cause a characteristic coarse (loose texture) facial appearance.
- *Hand and feet*—hand and feet become large, with clubbing of the toes and fingers due to enlargement of the tufts of the terminal phalanges. The terminal phalanges of the hands and feet become large and the ribs also increase in size.



Fig. 36-1: Increased size of skull and facial bone seen in gigantism.

Oral Manifestations

- *Symptoms*—in edentulous patients' enlargement of the alveolus may prevent the comfortable fit of complete dentures.
- *Teeth*—teeth in gigantism are proportional to the sized of jaw and the rest of the body and root may be longer than normal. The teeth become spaced, partly because of enlargement of the tongue and partly because upper teeth are situated on the inner aspect of the lower dental arch, due to disproportionate enlargement of the two jaws
- *Jaw bone*—mandibular condylar growth is very prominent. Overgrowth of mandible leading to prognathism.

Mandible may be of extraordinary proportions creating a major discrepancy between the upper and lower jaws and a pronounced class III malocclusion. In some cases the growth at the condyle exceeds that of the alveolar processes, so that increased in vertical depth of the ramus is greater than that of the body of the jaw, consequently the upper and lower teeth fail to come into proper occlusion.

- *Palate*—the palatal vault is usually flattened and the tongue increase in size and may cause crenation on its lateral border.
- *Macroglossia*—soft tissue growth may produce uniform macroglossia in patients.
- *Lips*—the lips become thick and Negroid.

Radiographic Features

- *Skull changes*—enlargement of sella tursica, enlargement of paranasal sinus (Fig. 36-2) and excessive pneumatization of temporal bone squames and petrous ridge. Diffuse thickening of outer table of skull. Enlargement and distortion of the pituitary fossa.

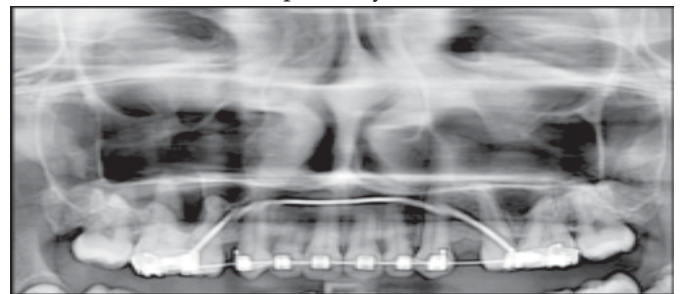


Fig. 36-2: Enlargement of maxillary sinus seen in hyperpituitarism patient.

- *Air sinus*—the air sinuses are really prominent in acromegaly rather than in gigantism.
- *Teeth*—increased tooth size especially root due to secondary cemental hyperplasia. Diastema between teeth due to lengthening of dental arch. Increase in thickness and height of alveolar process may results in open bite to the patient.
- *Jaw bone*—in acromegaly the angle between the ramus and body of mandible may increase, which result in anterior tooth root push forward so they appear as 'fan out'. There is also lengthening of condylar process, class III skeletal relationship. The new bone laid down on the condyle results in an increase in the vertical length of the ramus as well as overall length of whole bone. It is greater on the tip and posterior aspect of the coronoid processes, on the posterior border of the ramus and on the chin. Enlargement of the mandible, the length of the horizontal and ascending rami are both increase causing it to become prognathic with an increase obliquity of the angle and with loss of the antegonial notch.

- *Inferior dental canal*—there is also enlargement of inferior dental canal.

Diagnosis

- *Clinical diagnosis*—stature of individual gives clue to the diagnosis.
- *Radiological features*—characteristic radiographic findings may yield to the diagnosis.
- *Laboratory diagnosis*—growth hormone concentration can be measured by radioimmunoassay technique.

Management

- *Surgery*—trans-sphenoidal surgery may result in cure of GH excess especially in patients with macroadenoma.
- *Medical therapy*—octreotide, a long acting analogue of somatostatin, lowers GH. It is administered as subcutaneous insulin 2 to 3 times/day. Other somatostatin analogues like lanreotide, vapreotide can also be given in this patient. Dopamine antagonists are also used.
- *Radiotherapy*—external radiotherapy stops tumor growth and lowers GH levels. GH levels fall slowly and there is a risk of hypopituitarism.

Hypopituitarism or Pituitary Dwarfism

Pituitary is the master gland of the body. It results due to reduced secretion of pituitary hormone which may occur due to pituitary adenoma that compresses the pituitary gland. It is often the consequence of growth of pituitary gland or interference with stalk function. It results in *pituitary dwarfism*.

Pathologic changes can result from a variety of pituitary gland malfunction. Total absence of all pituitary secretions is known as *panhypopituitarism*. Hypopituitarism which commences after puberty is called as '*Simmond's disease*'.

Etiology

- *Disease of pituitary gland*—congenital or due to destructive disease of pituitary gland such as infarct occurring before puberty.
- *Space occupying lesion*—space occupying lesions involving the sella turcica like craniopharyngioma, adenomas and sarcoidosis.
- *Sheehan's syndrome*—Sheehan's syndrome is a form of hypopituitarism caused by infarction of the pituitary associated with postpartum hemorrhage.

Clinical Features

- *Short stature of individual*—the underdevelopment is symmetrical, individual is very small and in some cases there may be a disproportional shortening of the long

bones. The hallmark of this condition is that the growth is retarded to a greater degree than is bone and dental development (**Fig. 36-3**).

- *Hypocalcemia*—it may occur because of growth hormone and cortisol deficiency. Lack of gonadotrophin delays the onset of puberty.
- *Diabetic insipidus*—the presence of diabetes insipidus associated with deficient secretion of vasopressin is suggestive of pituitary dysfunction.
- *Symptoms*—growth hormone secretion is lost resulting in lethargy, muscle weakness and increase fat mass in adults.
- *Sexual characteristic*—luteinizing hormone (LH) secretion becomes impaired, in the male loss of libido and impotence and in female oligomenorrhea or amenorrhea. The male produces gynecomastia and skin becomes fair and wrinkled.
- *Skull*—the skull and facial bone are small and there is delay in maturation of the skeleton and epiphysis may remain ununited throughout the life.



Fig. 36-3: Short stature of individual in dwarfism.

Oral Manifestations

- *Jaw bone*—marked failure of development of maxilla and mandible with lack of condylar growth with short ramus and this can lead to severe malocclusion and crowding of the teeth.
- *Teeth*—the two important hormones are excreted by this gland—the somatotrophic and the thyrotrophic are responsible for the normal eruption of teeth and the

alveolar growth. Thus in case of hypofunction of this gland, the tooth eruption is hampered. The dental arch is smaller than normal and thus cannot accommodate all the teeth resulting in crowding and subsequent malocclusion. The clinical crown appears smaller than normal because even though eruption does occur it is not complete. Eruption is delayed and so the shedding of the deciduous teeth (Fig. 36-4).



Fig. 36-4: Malocclusion and delayed eruption of teeth seen in dwarfism patient

Radiographic Features

- *Teeth*—complete absence of third molar bud. Roots of teeth are short and apices are wide open and pulp canal toward the apex.
- *Alveolar bone*—there is loss of alveolar bone.

Diagnosis

- *Clinical diagnosis*—short stature of individual with respect to age will easily diagnose this condition. Eruption of teeth is also delayed.
- *Radiological diagnosis*—short root with wide open apices are present
- *Laboratory diagnosis*—radioimmunoassay will show level of growth hormone significantly below normal.

Management

- *Removal of cause*—management is usually directed towards removal of the cause.
- *Growth hormone replacement therapy*—this can be produced by recombinant DNA technology.

Progeria

It is transmitted as autosomal dominant trait. This condition is rare and underlying cause is entirely dependent on

pituitary dysfunction. It is regarded as premature senility in an individual of infantile proportions.

Clinical Features

- *Age*—affected infants appear normal at birth, but the typical clinical features become manifested within the first few years.
- *Symptoms*—patient exhibits alopecia, pigmented areas of the trunk, atrophic skin, prominent veins and loss of subcutaneous fat. The individual have high pitched squeaky voice beak like nose and hypoplastic mandible.
- *Sign*—the face is pointed, with the nose resembling the beak of a bird. The head is large, while mandible is small. Exophthalmos may be present and joint deformities. The lip is thin. The intelligence of this patient is either normal or above normal and even at early age patient behave like old person.

Oral Manifestations

- *Formation of irregular dentin*—there is accelerated formation of irregular dentin.
- *Delayed eruption*—delayed eruption of teeth can occur.

Radiographic Features

- *Long bone*—there is presence of osteoporosis of the long bones.
- *Skull bone*—there is relative overdevelopment of the frontal and parietal bones, while ossification may be delayed and deficient.
- *Jaw bone*—the mandible and maxilla are small, but the mandible shows the greatest amount of underdevelopment so that the chin is underhung.

Diagnosis

- *Clinical diagnosis*—alopecia, pigmented area on face, beak of bird nose, squeaky voice and delayed eruption of teeth may give clue to diagnosis.
- *Radiological diagnosis*—osteoporosis, delayed ossification, and underhung chin is present.

Management

No treatment and patient usually dies before the age of 27 years.

Diseases of Thyroid Gland

Hyperthyroidism

It is also called as 'thyrotoxicosis' and it is a syndrome in which there is excessive production of thyroxin in thyroid

gland. It is associated with diffuse toxic goiter and less frequently with toxic nodular goiter or toxic adenoma. Excessive thyroxine causes generalized increase in metabolic rate of all body tissues. In patient with thyrotoxicosis, dental treatment can precipitate an acute emergency like 'thyroid crisis' or 'thyroid storm'.

Etiology

- *Exophthalmic goiter*—it is characterized by diffuse hyperplasia of the thyroid and by eye signs.
- *Toxic adenoma*—in it hyperfunction originates by a benign tumor of the thyroid gland.
- *Pituitary disease*—a pituitary disease involving anterior portion of the gland.
- *Other causes*—ectopic thyroid tissue, Graves' disease, multi-nodular goiter, thyroid adenoma, choriocarcinoma, excess pituitary TSH, autonomous stroma ovarii, and polyostotic fibrous dysplasia.

Clinical Features

- *Age and sex*—it has predilection for females between 20 and 40 years of age.
- *Thyroid features*—thyroid is diffusely enlarged, smooth, possible asymmetrical and nodular, a thrill may be present, may be tender (**Fig. 36-5**).
- *Neuromuscular*—it includes nervousness, fine tremors, and muscle weakness, mood swings from depression to extreme euphoria, emotional lability, hyper-reflexia, ill sustained clonus, proximal myopathy, bulbar myopathy and periodic paralysis.
- *Gastrointestinal*—weight loss despite normal or increased appetite, diarrhea, bowel alterations, anorexia, vomiting and hyperdefecation. Abdomen, liver and spleen may be enlarged.



Fig. 36-5: Thyroid gland swelling seen in neck area due to tumor.

- *Cardiorespiratory*—palpitation, excessive perspiration, irregular heartbeat, increased metabolic activity leads to increased circulatory demands. There is also tachycardia, increased pulse pressure, and sometimes congestive cardiac failure. Exertion dyspnea, ankle edema, systolic hypertension, angina, and cardiomyopathy may be present.
- *Ocular*—in thyrotoxicosis patient may have bulging eye and partial paralysis of the ocular muscles, retraction and jerky movement, corneal ulceration, optic neuritis, ocular muscle weakness, papilledema, loss of visual activity, and exophthalmos is present.
- *Reproductive*—amenorrhea, oligomenorrhea, infertility, spontaneous abortion and loss of libido, impotence.
- *Dermatological*—increases sweating, pruritis, onycholysis, pigmentation, vitiligo, digital clubbing and pretibial myxedema (bilateral nonpitting edema).
- *Others*—heat intolerance, sweaty and warm extremities, thin shiny skin, pretibial myxedema, increased pulse rate and early fatigue, lymphadenopathy, thirst and osteoporosis.

Oral Manifestations

- *Teeth*—advanced rate of dental development and early eruption with premature loss of primary teeth.
- *Alveolar bone*—generalized decrease in bone density or loss of some areas of edentulous alveolar bone.

Radiographic Features

- *Generalized osteoporosis*—in older children and adults well marked generalized osteoporosis sometimes appears but it is not revealed in the jaw.
- *Alveolar resorption*—in some cases, there may be alveolar resorption and in some cases there may be greater density of the trabeculae.

Diagnosis

- *Clinical diagnosis*—enlargement of thyroid gland with other systemic features may give clue to diagnosis. Advanced rate of tooth development.
- *Radiological features*—generalized osteoporosis with alveolar resorption.
- *Laboratory diagnosis*—plasma levels of T_3 and T_4 are increased; free thyroxine index is raised in this disorder. Thyroid stimulating hormone (TSH) decreased. Anemia may be moderate to severe degree and is seen in patient with prolonged duration of the disease. The anemia is hypochromic and abnormal forms of RBC may be seen.

Management

- *General*—sedate the patient giving medication. Application of cold packs to lower the body temperature.

- *Antithyroid drugs*—it would be appropriated to give antithyroid drugs for 12 to 18 months to those in whom a single episode was anticipated. Carbimazole—for 0-3 weeks, 40-60 mg daily in divided doses; for 4-8 weeks, 20-40 mg daily in divided doses and for maintenance phase, 5-20 mg daily.
- *Subtotal thyroidectomy*—patients must rendered euthyroid before operation. The antithyroid drug is stopped 2 weeks before surgery and replaced by potassium iodate 170 mg daily orally.
- *Radioactive iodine*—¹³¹I acts either by destroying functional thyroid cells or by inhibiting their ability to replicate. Dose -185-370 mBq (5-10 mCi) is given orally.
- *β-adrenoreceptor antagonist*—selective β-adrenoreceptor antagonist such as propranolol (160 mg daily in divided doses or nadolol 40-80 mg once daily) will alleviate but not abolish symptoms of hyperthyroidism within 24-48 hours.

Hypothyroidism

It is caused by insufficient secretion of thyroxin by the thyroid gland. Failure of thyrotrophic function on the part of the pituitary gland or an atrophy or destruction of the thyroid gland leads to an inability of the thyroid to produce sufficient hormone to meet the requirement of the body. Hypothyroidism is caused by decreased or deficient secretion of thyroid hormones caused by thyroiditis, insufficient thyroid replacement, post-thyroidectomy, post-radioactive iodine therapy.

Types

- *Cretinism*—if failure of hormone occurs in infancy.
- *Juvenile myxedema*—if it occurs in childhood.
- *Myxedema*—if it occurs after the puberty. In it, there is subcutaneous deposition of glycosaminoglycan ground substance producing non-pitting edema.
- *Primary hypothyroidism*—thyroid gland is abnormal.
- *Secondary hypothyroidism*—in this pituitary gland does not produce adequate amount of thyroid stimulating hormone.

Clinical Features

Cretinism and juvenile myxedema

- *Age*—it may be present at birth or become evidence within the first few months after birth.
- *Symptoms*—hoarse cry, constipation, feeding problems in neonates. Retarded mental and physical growth. Patient also noticed huskiness of voice, constipation and hypothermia is present. Skin is dry to touch.
- *Bones*—delayed fusion of all body epiphysis and delayed ossification of paranasal sinus, partially pneumatization.
- *Signs*—there is protuberant abdomen with umbilical hernia. The hairs are sparse and brittle, the finger nails are brittle and the sweat glands are atrophic.

Myxedema

Symptoms

- *Early symptoms*—it includes weakness, fatigue, cold intolerance, lethargy, dryness of skin, headache, menorrhagia, and anorexia.
- *Late symptoms*—it includes slowing of intellectual and motor activity, absence of sweating, modest weight gain, constipation, peripheral edema, pallor, hoarseness, decreased sense of taste and smell, muscle cramps, aches and pains, dyspnea, anginal pain and deafness.

Signs

- *Face*—patient has dull expressionless face, periorbital edema, facial pallor, puffiness of face.
- *Eyes*—puffiness of eyelids is also present. There are also watery eyes.
- *Skin*—skin feels droughty to touch sparse hair
- *Tendon reflexes*—delayed return of deep tendon reflexes
- *Thyroid gland*—thyroid gland may be enlarged.
- *Other features*—patient may suffer from disorientation, tachycardia, diastolic hypertension, displaced apical beat, occasional purpura, thickened nose, and pleural effusion.

Complications

- Coronary artery disease, congestive heart failure.
- Increased susceptibility to infection, mental disturbances including depression.

Oral Manifestations

Cretinism and juvenile myxedema

- *Teeth*—dental development delayed and primary teeth slow to exfoliate. Enamel hypoplasia can also be seen. Abnormalities of dentin formation lead to enlarge pulp chamber.
- *Jaw bone*—maxilla is overdeveloped and mandible is underdeveloped. Retarded condylar growth leads to characteristic micrognathia and open bite relationship.
- *Tongue*—tongue is enlarged by edema fluid and due to it tongue may protruded continuously and such protrusion may lead to malocclusion of teeth.
- *Skull*—the base of skull is shortened leading to a retraction of the bridge of the nose with flaring.
- *Face*—face is wide and fails to develop in longitudinal direction.
- *Lips*—lips are puffy, thickened and protruding.

Myxedema

- *Tongue and lip*—macroglossia and enlarged lip as a result of the deposition of water and protein.
- *Face*—facial swelling of nonpitting type and mandible is underdeveloped.
- *Teeth*—there is greater tendency to periodontal disease, with alveolar destruction and loosening of the teeth.

Radiographic Features

- *Skull bones*—delayed closing of the fontanelles and epiphysis, numerous wormian bones (accessory bone in the sutures). There is transverse line of increased density involving the metaphysical regions.
- *Teeth*—reveal thinning of lamina dura. Delayed dental eruption, short tooth root. Separation of teeth as a result of enlargement of tongue, periodontal diseases and loss of teeth and external root resorption.
- *Alveolar bone*—the alveolar processes are relatively large as compared to body of bone which is smaller than in normal individuals.

Diagnosis

- *Clinical diagnosis*—sparse hair, delayed development of teeth, enamel hypoplasia and macroglossia should give clue to the diagnosis.
- *Radiological diagnosis*—thinning of lamina dura, wormian bone, and external root resorption can give clue to the diagnosis.
- *Laboratory diagnosis*—thyroid stimulating hormone (TSH) increased and T_3 and T_4 decreased. In ECG, a classical sinus bradycardia with low voltage complexes and ST/T wave abnormalities. There is raised cholesterol level and triglycerides level and low serum sodium.

Management

- *Thyroid preparation*—patients are managed by thyroid preparation. Mainly used is levothyroxin, which is available as 25, 50 and 100 mg tablets. It is customary to start slowly and a dose of 50 mg/day should be given for 3 weeks and finally to 150 mg/day. In the elders and in patient with ischemic heart disease, the initial dose should be 25 µg/day. In children it should start as early as possible to avoid major developmental and intellectual abnormalities.

Dental Consideration of Thyroid Disorders

- *Precaution*—in dentistry, the use of sedative and analgesic are dangerous as these agents tend to precipitate coma in patient with hypothyroidism.
- *Hyperparathyroidism*—inpatients with severe hyperthyroidism the emergencies likely to occur are thyroid crisis, emotional disturbances, cardiac difficulties.

- *Lowering the body temperature*—application of cold packs to lower body temperature
- *Other therapy*—other therapy like supporting respiration, narcotic antagonist, and oxygen can be given if necessary.
- *Hospitalization*—if severe, patient should be immediately hospitalized.

Diseases of Parathyroid Gland**Hyperparathyroidism**

It is an endocrine disorder in which there is an excess of circulating parathyroid hormone. Excess PTH stimulates osteoclast to mobilize calcium from skeleton leading to hypercalcemia in addition to PTH increased renal tubular re-absorption of calcium.

Following is the sequence of event which gives an idea of the reaction promoted by this hormone.

- *Target organ*—the bone and the kidney are the target organs of parathyroid hormone which mediates the osteoclast to resorb bone actively.
- *Elevation of serum calcium level*—when the bone is resorbed, calcium is released in the extracellular fluid and the serum calcium level is elevated.
- *Action of parathyroid hormone*—the parathyroid hormone acts on the epithelium of kidney tubules causing diuresis of phosphorus resulting in decrease in serum phosphorus level. At the same time it induces an increase in calcium re-absorption from glomerular filtrate. Parathyroid hormone may also increase the absorption of calcium from the intestine but this is not definitely established. Hence in a healthy person injection of parathyroid hormone produces an elevated plasma calcium level, a decreased plasma phosphorus level and an increased alkaline phosphatase level.

Types

- *Primary*—there is autonomous secretion of parathyroid hormone (PTH) by hyperplasia, benign and malignant tumor of one or more of the four parathyroid glands.
- *Secondary*—compensatory increase in output of PTH in response to hypocalcemia. The underlying hypocalcemia may result from an inadequate dietary intake or poor absorption of vitamin D or from deficient metabolism of vitamin D in the liver or kidney. Its effects to restore serum calcium level at the expense of the loss of calcium in bone.
- *Tertiary*—occasionally parathyroid tumor after long standing secondary hyperparathyroidism develops this condition known as tertiary hyperparathyroidism. The increased parathyroid level produces increased bone resorption and a resultant hypercalcemia.

- *Ectopic*—due to excessive parathyroid hormone synthesized in patient with malignant disease.

Clinical Features

- *Age and sex*—female to male ratio is 3:1. Mainly in 30 to 60 years of age.
- *Classic triad*—there is present of classic triad of kidney stones, resorption of bone and duodenal ulcers.
- *Renal calculi*—symptoms in patient occur due to renal calculi and duodenal ulcers. Patient may complain of hematuria, back pain, urinary tract infection.
- *Psychological problems*—patient may suffer from psychiatric effect like emotional instability.
- *Gastrointestinal problems*—gastrointestinal difficulties such as anorexia, nausea, vomiting and crampy pain may be present.
- *Bone*—bone pain, pathologic fractures and bone deformities occurs.
- *Hypercalcemia*—it is associated with muscle weakness, fatigue, weight loss, insomnia, headache, polydipsia and polyuria.

Oral Manifestations

- *Brown tumor*—it may develop peripherally or centrally. This can be presented as swelling which may appear intraorally or extraorally (**Figs 36-6 and 36-7**).
- *Teeth*—gradual loosening, drifting and loss of teeth, malocclusion.



Fig. 36-6: Extraoral swelling seen in patient with hyperparathyroidism due to brown tumor.

Radiographic Features

- *Demineralization of skeleton*—bone matrix contains less than normal amounts of calcium producing unusually



Fig. 36-7: Brown tumor presented as intraoral swelling in patient with drifting of teeth (Courtesy Dr Parate).

radiolucent skeletal image. There is lack of normal contrast in the radiograph resulting in over all grayness, often associated with a granular appearance in the bone.

- *Ground glass appearance*—the rarefaction is of homogeneous nature and there may be normal, granular or ground glass appearance.
- *Moth eaten appearance*—sometimes rarefaction gives a mottled or moth eaten appearance with varying density.
- *Osteitis fibrosa generalisata*—localized destruction of bone is produced by osteoclastic activity leaving residual area of fibrosis. In some cases moth eater appearance can be seen.
- *Brown tumor*—it appears radiographically as ill-defined radiolucency (**Figs 36-8 and 36-9**) called as brown tumor, as gross specimen is brown or reddish brown. In it,

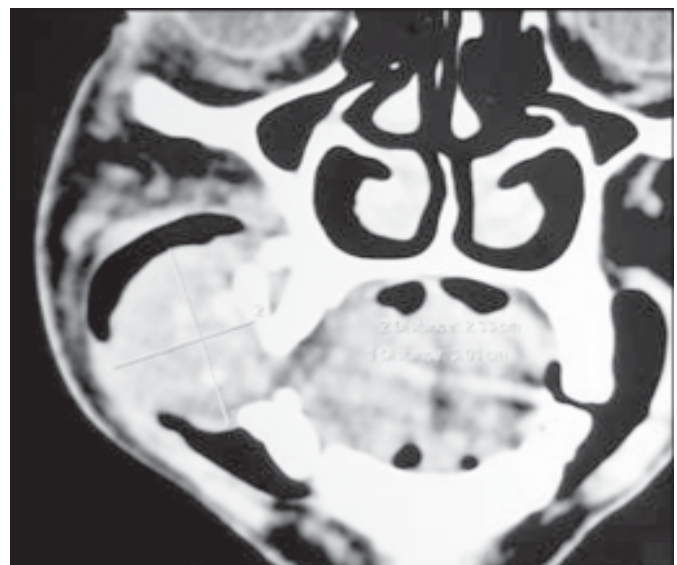


Fig. 36-8: CT scan of brown tumor in patient with hyperparathyroidism.



Fig. 36-9: Brown tumor presented as radiolucency in upper maxillary region. Note also there is drifting of teeth (Courtesy Dr Parate).

trabeculae are completely missing. It may occur in pelvis, ribs or femur but are most commonly found in facial bones and jaws. They appear as unilocular or multilocular with variably defined margin and may produce cortical expansion.

- *Pathological calcification*—punctuate and nodular calcifications occasionally occur in kidneys and joints.
- *Hand bone*—earliest change is subtle erosion of bone from sub-periosteal surface of phalanges of hand.
- *Skull bones*—entire calvarium has granular appearance caused by loss of central trabeculae and thinning of cortical tables.
- *Pepper pot skull*—evidence in the skull vault of osteopenia producing a fine overall stippled pattern to the bone, hence it is called as pepper-pot skull.
- *Jaw bones*—demineralization of inferior border of mandibular canal, thinning of outlines of the maxillary sinus.
- *Teeth and alveolar bone*—if the alveolus is severely affected the teeth may become mobile and migrate. The radiopaque cortical plate outlining the bones and anatomic regions may be thinned or lost entirely. Loss of lamina dura which may be seen around one tooth or all remaining teeth. It may be complete or partial. If complete involved tooth has tapered appearance.

Diagnosis

- *Clinical diagnosis*—brown tumor, symptoms of renal calculi, peptic ulcer can diagnose this condition.
- *Radiological features*—demineralization of skeleton, pepper pot skull, brown tumor, and osteitis fibrosa cystica can give clue to the diagnosis.
- *Laboratory diagnosis*—biopsy shows osteoclastic resorption of the trabeculae of the spongiosa and along the blood vessels in the haversian system of the cortex. Fibrosis especially in the marrow spaces is marked. The serum calcium level is raised and serum phosphorus

level is decreased and serum alkaline phosphatase level is elevated in primary hyperparathyroidism and in secondary hyperparathyroidism the serum calcium level is decreased whereas the serum phosphorus and alkaline phosphatase level are elevated. Increase in circulating hormone demonstrated by radioisotope studies.

Differential Diagnosis

Unilocular

- *Postextraction socket and surgical defect*—it will give history of extraction and surgery respectively.
- *Primordial bone cyst, traumatic bone cyst and odontogenic cyst*—they all occur in a younger age group than in hyperparathyroidism and have normal serum chemistry values.

Multilocular

- *Paget disease*—calcium metabolism is normal and bone formation usually exceeds the bone resorption resulting in an elevated blood alkaline phosphatase.
- *Ameloblastoma*—it usually shows a honeycomb appearance accompanied with paresthesia and normal serum chemistry level.
- *Central giant cell granuloma*—it can only be differentiated by serum chemistry level.
- *Cherubism*—the lesion is bilateral, seen in children and also there is usually familial involvement which is not so in giant cell lesion of hyperparathyroidism.
- *Aneurysmal bone cyst and central hemangioma*—it occurs in a younger age group.
- *Osteomalacia*—blood calcium levels are decreased.
- *Fibrous dysplasia*—osseous changes are frequently localized, loss of lamina dura is less common.
- *Multiple myeloma*—lesions are punched out, generalized bone demineralization in hyperparathyroidism helps in distinguishing.

Management

- *Surgery*—hyperplastic tissue should be removed surgically.
- *Vitamin D supplement*—the oral administration of vitamin D in secondary type can prevent skeletal demineralization in most of the cases.
- *Parathyroidectomy*—it is indicated in patient where patient does not respond to treatment.
- *Precaution*—restriction of dietary phosphate, phosphate binding agent and aluminum salts should be done.

Hypoparathyroidism

It is an uncommon condition in which there is insufficient secretion of parathyroid hormone. Parathyroid hormone is needed for regulation of calcium level in extracellular tissue.

Etiology

- *Surgical damage*—surgical damage to parathyroid gland and their vascular supply during thyroid gland procedure.
- *Damage from radiotherapy*—parathyroid damage may occur from radioactive iodine 131.
- *Autoimmune*—autoimmune destruction of parathyroid gland may take place.
- *Other*—hypoparathyroidism can also be associated with DiGeorge syndrome and endocrine-candidiasis syndrome.

Clinical Features

- *Hypocalcemia*—hypocalcemia can occur due to loss of parathyroid function. It can lead to tetany in the form of carpopedal spasm of the wrist and ankle joint.
- *Symptoms*—there is stiffness in hands, feet and lips. There is also paresthesia of hand, feet and around the mouth. Tingling in the circumoral area, fingers and toes. Patients may complaint of anxiety, depression, epilepsy and chorea. Reduction in intellectual capacity due to calcification within the brain.
- *Trousseau's sign*—it is elicited by occluding blood flow to the forearm for 3 minutes with sphygmomanometer cuff applied to the arm and raising the pressure above systolic level. This will induce carpopedal spasm.

Oral Manifestations

- *Teeth*—hypoplasia of enamel (**Fig. 36-10**), delayed eruption, external root resorption and root dilacerations. There is also blunting of molar roots.



Fig. 36-10: Enamel hypoplasia in patient with known history of hypoparathyroidism in childhood.

- *Chvostek sign*—a sharp tap over the facial nerve in front of ear causes muscle twitching of facial muscle around the mouth which is called as Chvostek sign.
- *Candidiasis*—chronic candidiasis is also some time present in case when hypoparathyroidism is associated with endocrine-candidiasis syndrome.

Radiographic Features

- *Calcification*—calcification of basal ganglion which appears flocculent and paired with the cerebral hemisphere on PA view.
- *Dental radiographic finding*—radiograph of jaw may reveal enamel hypoplasia, external root resorption, delayed eruption or root calcification.

Diagnosis

- *Clinical diagnosis*—Trousseau's sign, Chvostek sign and features of tetany may diagnose this condition.
- *Radiological features*—calcification of basal ganglion with enamel hypoplasia may give clue to diagnosis.
- *Laboratory diagnosis*—the serum calcium level is decreased usually below 7 mg/dl. Serum phosphate level correspondingly elevated. Urinary calcium is low or absent.

Management

- *Calcium and vitamin D supplement*—supplemental calcium and vitamin D depending on severity of the hypocalcemia and the nature of the associated signs and symptoms.
- *Intravenous calcium gluconate*—in severe cases intravenous administration of calcium gluconate is the treatment of choice.

Pseudohypoparathyroidism

It is also called as 'Albright Hereditary osteodystrophy', 'acrodysostosis'. In this normal parathyroid hormone is present in the body but biochemical pathway responsible for activating target cells are defective in function.

Types

- *Type I*—there are again three subcategories.
 - *Type I a*—molecular defect of intracellular binding protein prevent formation of cyclic adenosine monophosphate (cAMP). This will hamper cell metabolism. This is autosomal dominant.
 - *Type I b*—it is caused by defective receptor for the PTH on the surface of the target cells. It is autosomal dominant trait.
 - *Type I c*—there is defect in adenylate cyclase.

- *Type II*—there is induction of cAMP by PTH in target cells, but function response by the cell is not invoked.

Clinical Features

- *Stature*—patient manifest short stature due to early closure of certain bony epiphysis.
- *Shortened finger*—hand shows shortening of the metacarpal bones, so that finger are short.
- *Osteoma cutis*—subcutaneous calcification may be present in some patient

Oral Manifestation

- *Facial features*—midfacial hypoplasia, The face is rounded in appearance.
- *Teeth*—there is generalized enamel hypoplasia, oligodontia, and delayed eruption of teeth present.

Radiological Features

- *Apices of teeth*—there is blunting of apices of teeth
- *Dagger shaped calcification*—dagger shaped calcification seen in pulp of teeth.
- *Pulp chamber*—widened pulp chamber is present.

Diagnosis

- *Clinical diagnosis*—short stature, osteoma cutis oligodontia is present.
- *Radiological features*—dagger shaped calcification is typical of this disease.
- *Laboratory diagnosis*—elevated serum level of PTH with hypocalcemia, hyperphosphatemia.

Management

- *Vitamin D and calcium*—this can be given to control pseudohyperparathyroidism.

Diseases of Pancreatic Gland

Diabetes Mellitus

It may cause by autoimmune response. Principal laboratory sign are hyperglycemia. It is common endocrine disorders characterized by chronic hyperglycemia and abnormalities in carbohydrate and lipid mechanism. It is caused by disorders of carbohydrate mechanism resulting from insulin deficiency or ineffectiveness, producing hyperglycemia and glycosuria.

Types

- *Type I or Insulin dependent (IDDM)*—it occurs due to deficiency. There is lack of insulin production resulting in severe hyperglycemia and ketoacidosis.

- *Type II or non-insulin dependent (NIDDM)*—it occurs due to insulin resistance. Insulin resistance occurs in type II diabetes is due to an abnormal insulin molecule, an excessive amount of circulating antagonists and target tissue defect.

- Non-obese
- Obese
- Maturity onset diabetes of the young (MODY)

Pathogenesis

Type I diabetes mellitus

- *Increase blood glucose level*—as there is deficiency of insulin glucose will remain in blood as absorption of it is hampered. So blood glucose level is increased.
- *Glucose as main energy source*—as glucose is main source of energy in the body and it can not get absorbed patient feels tired and loosed its weight inspite increased food intake.
- *Polyuria and polydipsia*—due to hyperglycemia, osmolarity of blood and urine increased which will results in frequent urination which again lead to increase in water intake (polydipsia).

Type II diabetes mellitus

- *Decrease in number of insulin receptor*—in this condition there is decrease in number of insulin receptor results in non absorption of glucose in the body. Thus patient show insulin resistance.

Etiology

Type I diabetes mellitus

- *Viruses*—several viruses are been implicated including infection with mumps Coxsackie's B₄, retrovirus, rubella and cytomegalovirus and Epstein-Barr virus. Virus particle known to cause cytopathic or autoimmune damage to beta cells have been isolated from the pancreas.
- *Diet*—bovine serum albumin (BSA) a major constituent of cow's milk has been implicated in triggering type I diabetes. It has been shown that a child who has taken cow's milk early in infancy has been more prone to develop type I diabetes mellitus as compared to other who has taken breast milk.
- *Stress*—it may precipitate the development of type I diabetes by stimulating the secretion of counter regulatory hormones and possibly by modulating immune activity.
- *Immunological factors*—there is evidence that type I diabetes is a T cell mediated autoimmune disease. There is also HLA linked genetic predisposition. Monoclonal cell infiltration of pancreatic islets restoration in selective destruction of insulin secreting cells and induction of remission by immunosuppressive drugs such as cyclosporine suggest its immunological etiology.

Type II diabetes mellitus

- **Genetic**—the majority of cause of type II diabetes are multifactorial. Various types are associated with it like hepatocyte nuclear factor, glucokinase, and mitochondrial DNA and insulin receptors.
- **Environmental factors**
 - **Life style**—overeating, especially when combined with obesity and under activity is associated with developmental of type II diabetes.
 - **Malnutrition**—it is proposed that malnutrition *in utero* and the infancy may damage beta cell development at a critical period predisposing to type II diabetes later in life.
 - **Age**—age is an important risk factors for type II diabetes as it is principally disease of middle aged and elderly affecting 10% of the population over the age of 65.
 - **Pregnancy**—during normal pregnancy, insulin sensitivity is reduced through the action of placental hormone and this affect glucose tolerance. The term gestational diabetes refers to hyperglycemia occurring for the first time during pregnancy.

Clinical Features

- **Polydipsia**—there is excessive intake of fluid.
- **Polyuria**—there is excessive urine passage.
- **Polyphagia**—there is excessive hunger.
- **Breath**—there is presence of acetone breath.
- **Visual activity**—visual difficulty ranging from progressive color blindness to total blindness that have disease more than 20 years.
- **Atherosclerosis**—coronary artery disease and stroke are frequent complication.
- **Diabetic neuropathy**—due to atherosclerosis nerves may get compressed causing marked irritability.
- **Infection**—recurrent vaginal (yeast) infections, recurrent urinary tract infections, recurrent skin infections (especially of feet) and reversible paraesthesia of fingers or toe.
- **Others symptoms**—patient may complaint of nocturia, weight loss, fatigue, obesity, temperature, and hypertension and reduced peripheral pulses.
- **Symptoms of type II diabetes mellitus**—symptoms are very mild in nature with this disease discovered on routine hematological examination.

Oral Manifestations

- **Effect on periodontium**—it will influence the onset and course of periodontal disease. Patient with diabetes are more prone to develop periodontal disease than are those with normal glucose metabolism. As such diabetes mellitus does not cause periodontal disease directly but

it alters the response of the periodontal lesion to local irritants, hastening bone loss and retarding postsurgical healing of the periodontal lesions. Also gingival fluid in the diabetes has more glucose level which favors the growth of microflora.

- **Periodontitis**—the patient may exhibit a fulminating periodontitis (**Fig. 36-11**) with periodontal abscess formation. This will give rise to mobility of teeth. There is severe and rapid alveolar bone resorption takes place. Insulin dependent diabetic children tend to have more destruction around the first molars and incisors than elsewhere.



Fig. 36-11: Severe periodontitis seen in diabetic mellitus patient of type I (Courtesy Dr Soni).

- **Median rhomboidal glossitis**—diabetes is considered to be factor for median rhomboidal glossitis as frequency of abnormal blood sugar level in diabetes is related to occurrence of median rhomboidal glossitis. There is also impairment of blood supply to dorsum of tongue due to arteriosclerotic changes in the blood vessels supplying the area. Impairment of local immune mechanism which decrease the concentration of Langerhans of cells in the lesion.
- **Oral candidiasis**—it is the infection with candida albicans which is occur due to encouragement of local multiplication of candida albicans due to impaired glucose level and immune mechanism.
- **Localized osteitis**—dry socket develops in diabetes hence they show delayed healing and impaired immunological balance.
- **Burning mouth**—it is often associated with variety of otherwise unexplained oral symptoms such as burning sensation, atypical paresis, dysesthesia and dysgeusia.
- **Trigeminal nerve involvement**—diabetes neuropathy is recognized as polymorphic condition as when manifested as polyneuropathy on the assumption that trigeminal nerve might be involved.

- *Xerostomia and increase caries activity*—there is also increased caries activity which results of xerostomia which occur due to excessive fluid loss.
- *Delayed healing*—there is delay in healing of oral wound due to decreased polymorphonuclear chemotaxis.
- *Diabetic sialadenosis*—in diabetic patient there is also diffuse nontender bilateral enlargement of parotid gland.
- *Other features*—there is also angular cheilosis, altered taste sensation, oral lichen planus, and diffuse enlargement of parotid gland.

Radiographic Features

- *Loss of lamina dura with blurring of alveolar crest*—slight discontinuity or blurring of the cortex of alveolar crest to wide destruction of lamina dura.
- *Bone loss*—there is also horizontal and vertical bone loss.

Complications

- *Microangiopathy*—it results in occlusion of small blood vessels producing peripheral vascular disease. This results in decreased tissue perfusion which in turn results in severe infection like gangrene.
- *Coronary artery disease*—vascular occlusion may affect the coronary artery resulting in myocardial infarction, cerebrovascular accident and stroke.
- *Blindness*—when retinal vessels are affected blindness may result.
- *Other complication*—ketoacidosis, premature mortality, and diabetic coma may result.

Diagnosis

- *Clinical diagnosis*—polyuria, polydipsia and polyphagia with periodontal problems may give clue to the diagnosis.
- *Radiological diagnosis*—not specific
- *Plasma glucose concentration*—unequivocal elevation of plasma glucose concentration greater than 140 mg/dl.
- *The glucose tolerance test*—plasma glucose concentration is 200 mg/dl.
- *Taste paper strip*—strips are available for direct estimation of blood glucose level.
- *Blood*—random glucose elevated, fasting glucose elevated, 2 hour postprandial glucose elevated.

Management

- *Diet control*
 - *Balance calorie intake*—the patient with NIDDM, particularly those who are obese, dietary control toward a balanced calorie intake and exercise leading to weight loss is the sole treatment require.
 - *Calories from carbohydrate*—in diabetic patient, it is recommended that the percentage of calories derives

from carbohydrates should be increase and that from fat reduced. A suitable diet for diabetic person—50% of the daily caloric intake should be derived from carbohydrates of which significant amounts should be in the form of non-starch polysaccharides.

- *Mono unsaturated oil*—fat intake should be reduced; the use of mono unsaturated oils in the diet (e.g. olive oil, peanut oil) is beneficial.
- *Other*—reduced intake of sodium (no more than 6 g/day), alcohol abstinence, low calories and sugar free drink are useful for patients with diabetes.
- *Oral hypoglycemic drugs*—if dietary management proves ineffective in controlling hyperglycemia, hypoglycemic drugs like insulin or oral hypoglycemic are prescribe.
 - *Sulfonylurea*—first generation sulfonylurea in which tolbutamide is nowadays is given in a dosage of 25 or 500 mg 8 or 12 hourly. It is useful in the elderly in whom the risk and the consequence of inducing hypoglycemia are increased. In second generation sulfonylurea, gliclazide and glipizide are widely used.
 - *Biguanides*—they are less widely used than sulfonylurea because of their higher incidence of side effects. Mechanism of action is by increasing insulin sensitivity and peripheral blood glucose uptake. Metformin in dose of 500 mg 12 hourly. Its use is contraindicated in persons with impaired renal or hepatic function and in those who take alcohol in excess because of risk of lactic acidosis.
 - *Alfa-glucosidase inhibitors*—they delay carbohydrates absorption in the gut by selectively inhibiting disaccharides. Acarbose is currently available in a dose of 50-100 mg with each meal.
- *Insulin therapy*—insulin was discovered in 1921. The duration of action of short acting unmodified insulin, which is a clear solution can be extended by addition of protamine and zinc at neural pH. Insulin is injected subcutaneously into recommended sites namely anterior abdominal wall, upper arms, outer thighs and buttocks. Short acting insulin has to be injected at least 30 minutes before a meal to allow adequate time for absorption. After giving insulin carbohydrate should be given to patient as blood glucose level may fall drastically and insulin shock may results. Side effects of insulin are hypoglycemia, weight gain, peripheral edema, insulin antibodies, local allergy and lipodystrophy.

Diabetic Insipidus

- *Causes*—it occurs due to insufficiency of the posterior pituitary hormone. Traumatic episodes, such as head trauma or surgical procedures carried out near the

pituitary region, can often lead to destruction of the posterior lobe of the pituitary. In these patients, there is damage to the neurohypophyseal mechanism for the production of vasopressin. Other causes of posterior pituitary disorders leading to diabetes insipidus include tumors like craniopharyngioma, syphilis and basal meningitis.

- **Symptoms**—there is increase thirst and passage of large quantities of urine. Urine is of low specific gravity. There is also dehydration, headache, irritability, and fatigue may be cause of restriction of fluid.
- **Management**—administration of vasopressin is the treatment of choice. Desmopressin can be given intranasal in a dose of 5-10 mg once or twice daily.

Dental Consideration

- **Emergency**—emergency can occur due to diabetes coma or insulin shock. If tolbutamide, chlorpropamide or small doses of insulin is taken by the patient there will be less chances of diabetic coma. However, if the patient takes large daily dose of insulin there is possibility of diabetic coma or insulin shock. If the patient complains of being thirsty, nauseous and shortness of breath and has warm dry skin, the patient is most likely hyperglycemic and should be immediately refer to the physician. No treatment will be required by the dentist.
- **Dental management**—deliver treatment in such a way so as to minimize disturbances of metabolic balance.
 - **Appointment**—appointment should be of short duration and in the morning. Encourage to maintain their standard regimen.
 - **Glucose drink**—glucose drink should be available if patient complains of hypoglycemia.
 - **Local anesthesia without epinephrine**—use local anesthesia without epinephrine in the dental procedure. There is increased incidence of dry socket is due to decreased blood supply to mandible is caused by arteriosclerosis in long standing diabetes. If you are giving epinephrine, it will further reduce blood supply which further increases chances of dry socket.
 - **Suturing**—following extraction suturing of the socket should be done to aid homeostasis.
 - **Physician referral**—physician advice should be taken before arranging general anesthesia for dental treatment.
 - **Antibiotics prophylaxis**—antibiotic prophylaxis before dental surgery to prevent subsequent infection.
 - **Avoid complicated oral procedure**—complicated oral procedure in dental emergencies should be avoided whenever possible in uncontrolled diabetics until stabilization of blood glucose level is achieved.

Diseases of Adrenal Gland

Addison's Disease

It is also called as '*chronic adrenal insufficiency of the adrenal cortex*'. It was first described by Addison in 1855.

Etiology

- **Autoimmune**—sporadic and polyglandular syndrome may cause destruction of adrenal gland. It may be cause by bilateral destruction of the suprarenal glands.
- **Infection**—tuberculosis and some deep fungal infection may cause this disease.
- **Other cause**—metastatic carcinoma, intradermal hemorrhage, amyloidosis, hemochromatosis, adrenal infarction and congenital adrenal hypoplasia may be the causative factors.
- **Drugs** which can cause Addison's disease are aminoglutethimide, ketoconazole and etomidate.

Clinical Features

- **Age and sex**—it is more common in males and, while found in all age groups, it is most frequently seen in the 3rd and 4th decade.
- **Symptoms**—feeble heart action, general debility, vomiting, and diarrhea and severe anemia. Patient complains of postural hypotension. There is also reduced resistance to infection, trauma, and stress.
- **Sign**—the disease is characterized by bronzing of skin, a pigmentation of the mucous membrane.
- **Metabolic function**—decrease cortisol level interferes with the manufacture of carbohydrates from protein, causing hypoglycemia and diminished glycogen storage in the liver.
- **Neuromuscular function**—neuromuscular function is inhibited, producing muscle weakness.

Oral Manifestation

- **Bronze pigmentation**—the pale brown or deep chocolate pigmentation of the oral mucosa, spreading over the buccal mucosa from the angle of the mouth and/or developing on the gingiva, tongue (**Fig. 36-12**), and lips may be first evidence of disease.

Diagnosis

- **Clinical diagnosis**—bronze skin and bronze pigmentation in oral cavity, decreased metabolic function may give clue to the diagnosis.
- **Laboratory diagnosis**—biopsy of oral lesion show acanthosis with silver positive granules in the cells of the stratum germinativum. Anemia is normocytic and normochromic associated with reticulocytosis. High blood levels potassium and low concentration of sodium and chloride. There is also elevated blood urea nitrogen



Fig. 36-12: Diffuse pigmentation seen on tongue in patient with Addison's disease.

Management

- *Glucocorticoids replacement*—cortisol is the drug of choice. In patient who are not critically ill hydrocortisone 15 mg on waking and 5 mg at 6 pm in evening.
- *Supplement Mineralocorticoids*—it can also be given.

Adrenogenital Syndrome

It refers to any situation in which there is overproduction of androgens. It results when hyperplasia or tumors of the adrenal cortex occur. It may appear at 3 different times of life, i.e. at birth, in childhood and in adult. Clinical features vary according to appearance of lesion.

- *At birth*—in female child it produces pseudohermaphroditism, while in male child it produces macrogynitosomia praecox.
- *In childhood*—in the females it produces masculinization and in males it produces sexual precocity.
- *In adults*—in females it produces virilism and in males it produces feminization.

If the disease begins early premature eruption of the teeth may occur. This disease managed by administration of corticosteroid or estrogen.

Cushing's Syndrome

Cushing's syndrome arises from excess secretion of glucocorticoids by the adrenal glands. It is described by Harvey Cushing in 1932.

Etiology

- *Adrenal tumor*—Cushing's syndrome is caused by adrenal adenoma, adrenal carcinoma, adrenal hyperplasia and basophilic adenoma of the anterior lobe of pituitary gland.

It can also be caused by ACTH secreting tumor of the anterior pituitary which is associated with adrenal cortical hyperplasia.

- *Administration of corticosteroid*—in some cases administration of exogenous corticosteroid may lead to this condition.
- *Ectopic*—ectopically located adrenal like tumor, e.g. in ovary may cause these conditions.
- *Other*—alcohol excess, major depressive illness and primary obesity.

Clinical Features

- *Age and sex*—female to male ratio is 3:5, seen in 3rd and 4th decades.
- *Symptoms*—patient may complain of weight loss, menstrual irregularity, hirsutism, backache, obesity, and hypertension.
- *Moon face*—rapidly acquired obesity about upper portion of the body due to abnormal deposition of fat will result in moon facies (Fig. 36-13).



Fig. 36-13: Rounded moon facies seen in Cushing's syndrome (Courtesy Dr Milind Chandurkar).

- *Buffalo hump*—there is truncal obesity with prominent supra-clavicular and dorsal cervical fat pads giving rise to the 'buffalo hump' appearance at the base of neck.
- *Purple abdominal striae*—dusky plethoric appearances with formation of purple striae appear on abdomen (Fig. 36-14).
- *Others feature*—thin distal extremities, concurrent diabetes and alternation in hair distribution can also be present.

Oral Manifestations

- *Dental age*—in children growth and development including skeletal and dental age may be retarded.



Fig. 36-14: Purple abdominal striae seen on abdomen in Cushing's syndrome (Courtesy Dr Milind Chandurkar).

Radiographic Features

- *Osteoporosis*—generalized osteoporosis is present. The bone likely to involve are the vertebrae and the ribs although the long bones may be affected.
- *Demineralization*—there is osseous demineralization with pathological fractures, often superimposed.
- *Skull*—it may show diffuse thinning and have mottled appearance.
- *Lamina dura*—jaw may show areas of loss of lamina dura.

Diagnosis

- *Clinical diagnosis*—moon facies, buffalo hump, purple striae on abdomen will easily diagnosed this condition.
- *Radiological features*—generalized osteoporosis with loss of lamina dura may give clue to diagnosis.
- *Laboratory diagnosis*—this is done by measuring serum ACTH and cortisol level after administration of dexamethasone. In normal patient this level will be decreased but it is unaffected in patient with Cushing's syndrome.

Management

- *Surgery*—tumors involving the adrenal cortex are removed surgically and often require postoperative administration of corticosteroids to maintain normal glucocorticosteroids level.
- *Radiotherapy*—in case of Cushing's syndrome due to adrenocortical hyperplasia, pituitary irradiation is the best treatment.
- *Drugs used*—metyrapone in dose of 2-6 gm per day in divided doses by mouth. Other drug given is *aminoglutethimide*, *ketoconazole* which act by blocking steroid synthesis.

- *Prevention*—when corticosteroid is prescribed lowest dose should give to patient to manage the immunological disease.

Adrenal Insufficiency

It is relatively rare and usually occurs in connection with an acute septicemia and is called as *Waterhouse-Friderichsen syndrome*. This occurs in patient who are on long-term steroid for the treatment of variety of systemic condition developed adrenal insufficiency.

Types

- *Primary*—it occurs due to disorders of pituitary or adrenal glands (primary adrenal insufficiency is called as Addison's diseases).
- *Secondary*—it occurs due to chronic administration of corticosteroid resulting in the suppression of endogenous steroid.

Etiopathogenesis

- *Sudden withdrawal of steroid*—it usually occur following sudden withdrawal of steroid hormones in a patient who has primary adrenal insufficiency. Following sudden withdrawal in a patient with normal adrenal cortices but with a temporary insufficiency resulting from cortical suppression by exogenous corticosteroid administration
- *Following stress*—following stress such as physiologic or psychological stress.
- *Bilateral adrenalectomy*—following bilateral adrenalectomy or removing of a functioning adrenal tumor that had been suppressing the other adrenal gland.
- *Destruction of pituitary gland*—following sudden destruction of the pituitary gland.
- *Trauma*—following injury to both adrenal glands by trauma, hemorrhage, infection, thrombosis or tumor.

Clinical Features

- *Age*—it occur primarily in children but can occur in adults.
- *Onset*—it is characterized by rapidly fulminating septic course, a pronounced purpura and death within 48 to 72 hours.
- *Symptoms*—in this, patient is not able to tolerate the stress. There is anxiety, fatigue, hypotension, abdominal pain, nausea, vomiting, cold clammy skin, lethargy, and partial or complete loss of consciousness. There is also mental confusion occur in acute adrenal insufficiency.
- *Sign*—oral, conjunctival, and vaginal mucosae often show patches of pigmentation. In some patient, particularly in the dark-skinned races, patchy area of

depigmentation surrounded by hyperpigmentation may be seen. Hyperpigmentation of buccal or labial mucosa is seen in many patients. These patches may be brown, gray or blue in color.

Oral Manifestations

- *Early eruption*—teeth may erupt early, compared with the normal, but the eruption is in harmony with the skeletal age.

Diagnosis

- *Clinical diagnosis*—stress intolerance, mental confusion and hyperpigmentation of buccal and labial mucosa will give clue to the diagnosis.
- *Laboratory diagnosis*—plasma cortisol levels are low and fail to raise after administration of ACTH. Plasma ACTH levels are elevated and serum sodium and chloride and urinary 17-ketosteroid and 17-hydroxy-corticosteroids are low.

Management

- *Replacement therapy*—it is given in combination of glucocorticoids (cortisol), mineralocorticoids (fludrocortisone), and anabolic steroids. In mild case, hydrocortisone alone might be sufficient.

Dental Considerations

- *Defer the treatment*—if the adrenal crisis occurs during the treatment further dental treatment should be stopped.
- *Position of patient*—put patient in shock position.
- *Airway maintenance*—maintain the patency of the airway and administer oxygen.
- *Physician consultation*—you should call physician for further management.
- *Saline*—start intravenous 5% dextrose saline.
- *Hydrocortisone*—administer 100-200 mg of hydrocortisone.
- *Sodium succinate*—sodium succinate intravenously should be given.

Prevention

- *History*—a careful history should be taken if the patient has been using steroids on long-term basis.
- *Additional steroid dose*—such patients should be administered additional dose of steroids prior to surgery such that dose is increased to double or triple level 2 to 3 days preceding the surgery. Then it should be gradually taper to the maintain level. Patient who has been on steroid therapy but presently is off this therapy for the last one year without any adverse effects, needs no steroid supplementation.

- *Physician consultation*—consult the physician to determine the suitable dose during the pre, intra, and postoperative phase.

Diseases of Gonads

Hypergonadism

If occurring in children, results in precocious puberty. The long bone develops quickly and child may initially turn toward tallness, but this is offset by the early fusion of the epiphyses so that adult's person may be short.

Hypogonadism

It occurs in equal frequency in males and females. The bones are long and slender and epiphyses are late in fusion. The supraciliary ridges, malar bone and the mandible show greater development.

The chin is pointed, the palate is high and markedly arched and irregularities of the teeth occur. The mandible tends to become enlarged and even massive with short rami in male hypogonadism.

The skull is small and there is marked or even excessive enlargement of the frontal and sphenoid sinuses and especially mastoid air sinus.

Pregnancy

Pregnancy cause physiologic changes throughout that are of relevance to dentist. Alteration in the level of circulating female sex hormone modified the response of periodontium. During pregnancy there are carious hormonal change leading to an increased blood volume, cardiac output, glomerular filtration rate and O₂ requirement.

- *First trimester*—during 1st trimester, there is persistence nausea and vomiting and morning sickness due to carbohydrate starvation and ketosis.
- *Third trimester*—during 3rd trimester as a response to emotional and physical stress of the treatment, large quantities of the steroid are liberated into bloodstream. The pituitary secretes oxytocin which can stimulate uterine contraction and hence premature labor.
- *Pregnancy gingivitis*—One of the most common oral complications seen with pregnant women is called pregnancy gingivitis. This condition presents as fiery red swollen gums. It usually begins in the second month of pregnancy and progresses throughout the pregnancy. It is the result of an exaggerated inflammatory response of the gums to bacteria, plaque and calculus in the mouth, due to an increase of estrogen and progesterone hormones in the pregnant woman. In pregnancy gingivitis, like regular gingivitis, the gum

tissue may bleed more easily than normal. After pregnancy these conditions should return to normal with proper oral hygiene.

- **Diabetes**—women can get gestational diabetes (hyperglycemia occurring for first time during pregnancy). Repeated pregnancy can increase likelihood of developing permanent diabetes particularly in obese women. Pregnancy in diabetic women is associated with increased perinatal mortality rate.
- **Epilepsy**—epilepsy may worsen during the pregnancy particularly during the 3rd trimester when plasma anti-convulsant levels tend to fall frequently, monitor blood level during pregnancy.

Dental Management

- **Routine dental procedure**—scaling and prophylaxis should be performed as often as is necessary to control local etiologic factors and reduce gingival inflammation. Second trimester is the safest period during which to perform routine dental care. Stressful procedure should be avoided during pregnancy. In a pregnant woman stress and strain involved can lead to complication during the treatment.
- **Radiograph**—dental radiographs or X-rays are not used for routine dental care. In emergency situations, X-rays may be needed to diagnose a problem, however proper safety measures should be taken. These measures include the lead apron, reduced radiation and the X-ray beam position. This again is most important in the first trimester.
- **Drugs**—we must also modify drugs that can be administered to the pregnant or breastfeeding patient. Certain anesthetics, pain relievers, antibiotics, and sedative drugs that may be used are considered unsafe for the developing fetus and must be avoided. For breastfeeding mothers there is concern over certain drugs that may be absorbed into the mother's milk. The local anesthetic that we use is considered safe for emergency dental care needs.
- **Syncope**—if syncope develops turning the patient on left side will relieve the pressure and put the patient back to normal. Supine position can lead pressure on inferior vena cava by the fetus leading to poor venous return and lead to hypotension further aggravating the condition.
- **First and third trimester**—all planned surgical procedures should be avoided in the 1st and 3rd trimester to avoid fetal stress and premature labor. This is because of the vulnerability of the growing fetus in the first 3 months or trimester and the comfort level of the mother in the last trimester. Third trimester dental care is best postponed due to the comfort level of the patient and to avoid supine

hypotension syndrome. This is a compression of the large vein in the abdomen, the inferior vena cava, which brings blood back to the heart. Unfortunately, lying on your back in the dental chair can cause this compression. Also, there is always the risk of the patient going into labor and dentists are not very good at delivering babies.

- **Precaution**—the best measure that the pregnant women can take is prevention. This consists of diligent brushing and flossing to remove plaque from the teeth and gums along with annual dental check-ups and cleanings. By maintaining a healthy oral environment, you can minimize the need for emergency dental treatment during your pregnancy.

Menopause

It begins when menstrual function ceases and it usually occur between the ages 40 to 55. There are numerous symptoms associated with it and they are caused by deficiency of estrogen.

Irritability, insomnia, nervousness, osteoporosis, back and joint pain can also occur in menopause. Patient may complaint of burning mouth and tongue, taste abnormalities and dryness of the mucous membrane are also present. There may be desquamative gingivitis in which there is atrophy and ulceration of the gingival tissue and bleeding.

Treatment consists of estrogen replacement therapy depending on the symptoms.

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Introduction

The cellular elements of the blood as well as its plasma proteins play an extraordinary role in many physiologic mechanisms of the body. The circulating blood has a profound effect on the maintenance of normal oral mucous membrane, especially in the mucosa of tongue.

Many important disorders of blood present with symptoms and signs are seen in the oral cavity. The dentist is frequently the first medical personnel to encounter such disorders.

Physiology of Blood

Blood is unique bright red (arterial) to dark red (venous) colored liquid of variable composition, circulating through the vessels of body.

It participates in all physiologic and pathologic activities in all organs and is composed of liquid called as '*plasma*' in which there are suspended erythrocytes, leukocytes and thrombocytes collectively called as '*hemocytes*'.

If blood is allowed to clot, an amber colored liquid remains after the separation of the clot and is known as '*serum*'. It differs from plasma only by the loss of protein fibrinogen, which is removed in the coagulation process.

Composition

Blood is a highly complex fluid, which is composed of two parts liquid plasma (55%) and different types of cells (45%).

- *Cells*—it consists of red blood cells or corpuscles or erythrocytes, white blood corpuscles or leukocytes and platelets or thrombocytes
- *Water*—it is about 91 to 92%.
- *Solid*—it consists of sodium, potassium, calcium, magnesium, phosphorus, iron, copper, etc.—0.9%.

Serum albumin, serum globulin, fibrinogen—7.5%. Nonprotein nitrogenous substances like urea, uric acid, carbohydrates, fats, etc. Various enzymes like amylase, protease and lipase are also present. The yellow color of plasma is due to bilirubin and carotene.

Plasma Proteins

In normal circumstances, total amount of plasma proteins varies from 6.5 to 7.5% and an average of 7%.

Composition

- *Serum albumin*—4.7 to 5.7%
- *Serum globulin*—1.8 to 2.5%
- *Fibrinogen*—0.2 to 0.4%
- *Prothrombin*—0.1 %

Functions of Plasma Proteins

- *Blood clotting*—they are essential for blood clotting.
- *Antibodies*—the antibodies are globulin in nature and are essential for defence against infection.
- *Transport*—they also help in transport of certain substances in blood like hormones and enzymes.
- *Osmotic pressure*—it maintains colloidal osmotic pressure of blood and concerned with erythrocyte sedimentation rate (ESR).
- *Buffer*—they act as a buffer in maintaining acid-base balance, as a protein reservoir.
- *Trephones*—the leukocytes prepare substances from plasma proteins which are necessary for nourishment of tissue cells.

Some Facts about Blood

- *Blood volume*—the total blood volume in the circulation as well as blood in stores is about 5 liters or 90 ml per kg of body weight.

- *Specific gravity of blood*—1.052 to 1.063 and primarily depends upon the ratio of plasma to red cells.
- *Viscosity of blood*—the relative viscosity of water, plasma and whole blood are roughly 1, 1.8 and 4.7.
- *pH*—the pH of blood varies between 7.36 and 7.45, the average is about 7.4.

Red Blood Corpuscles (Erythrocytes)

It was first described by Dutch microscopist *Leeuwenhoek* in 1674. It lacks cytoplasmic organelles such as nucleolus, mitochondria and ribosomes. It is unable to synthesize new proteins and carry out the oxidative mechanisms associated with mitochondria or undergo mitosis. They are produced in liver, spleen and lymph nodes in embryonic life and after birth; they are produced exclusively in the bone marrow.

Factors Controlling Erythropoiesis

- *Diet*—food rich in first class proteins is important as they supply essential amino acids for the synthesis of globulin for hemoglobin.
- *Tissue oxygenation*—any condition leading to decreased oxygen in the tissues increases the rate of RBC production, e.g. anemia, high altitude, prolonged cardiac failure and lung disease.
- *Erythropoietin stimulating factor*—it is a circulating hormone which is formed in the kidney.
- *Vitamins*—vitamin B₁₂ which is required for DNA synthesis and folic acid which is required for conversion of pre-erythroblast into early normoblast. Vitamin C and B₆ are also important factors.
- *Metals*—iron for hemoglobin formation; copper, magnesium and cobalt for conversion of iron into hemoglobin by catalytic action and calcium which help indirectly by conserving iron and its subsequent assimilation.
- *Bile salts*—they are required for proper absorption of metals.

Composition and Morphology of RBC

- Each cell is composed of a colorless envelope consisting of 65% water and 35% solids of which 33% is hemoglobin bound to 2% stromal network.
- *Shape and size*—normal RBC is biconcave disc having a mean diameter of approximately 6.9-7.4 microns. The shape of RBCs can change remarkably as the cell passes through the capillaries.
- *RBC count*—the normal RBC count in an adult male is taken as 5 to 5.4 million/mm³ cells per cu mm and in adult females, 4.5 to 4.8 million/mm³. In infants, the count is 6 to 7 millions/mm³ where as in fetus it is 7 to 8 million/mm³.

- *Variations in RBC count occur in the following physiological conditions*—the count is lowest during sleep, then gradually rises and becomes maximum in the evening. Also muscular exercise, high altitude and high external temperature lead to reduced oxygen tension in arterial blood; injection of adrenaline and excitement increase the RBC count.
- *Life span*—the average life span of a mature red cell is about 120 days. As the cells become senile, they become flask-shaped and become more brittle, then get fragmented and are swallowed up by the RE cells. Hemoglobin is released and broken down into heme and protein.

Functions of RBC

- *Respiratory*—they carry oxygen and carbon dioxide.
- *Acid base balance*—they help to maintain the acid base balance.
- *Viscosity*—they maintain the viscosity of blood.
- *Pigment*—various pigments are derived from hemoglobin after disintegration of RBCs.

Hemoglobin

It is a red conjugated protein. The hemoglobin molecule is roughly spherical in shape, consisting of two pairs of polypeptide chains to which a highly colored heme group, which is a complex of iron and protoporphyrin, is added. The protein portion of the molecule is called globin.

Functions of Hemoglobin

- *Oxygen carriage*—it is essential for oxygen carriage. It combines easily with oxygen to form oxyhemoglobin. Oxyhemoglobin holds oxygen loosely which can be easily displaced by many other gases like CO, CO₂, H₂S, etc. to form a more stable compound.
- *Carbon dioxide transport*—it helps in carbon dioxide transport by directly combining with CO₂ to form a carbamino compound.
- *Acid base balance*—it constitutes one of the important constituents of blood and helps in maintaining acid base balance of blood.
- *Pigment*—various pigments of bile, stool, urine, etc. are formed from it.

White Blood Corpuscles (Leukocytes)

They are the mobile units of the body's defence system. They are formed partially in the bone marrow and partially in the lymph tissue. After formation, the blood and the lymphatics transport them to different parts of the body. They are nucleated cells with hemoglobin. Leukocytes are rich in nucleoproteins and contain lipids, glycogen,

cholesterol, ascorbic acid and a variety of enzymes, especially proteolytic.

The average total number of WBCs, 6000 to 8000 cells per cu mm, the normal range being 4000 to 11000 cells per cu mm. Life span of WBC is about 2-4 days to about 12-15 days.

Physiologic Variations

Physiologic variations in WBC count occur due to

- *Diurnal variation*—in the morning or after rest the count is the lowest, which rises after mid-day and usually becomes highest in the evening.
- *Exercise*—muscular exercise and injection of adrenaline increases the count.
- *Age*—in a newborn, the count is very high, i.e. about 20,000 cells per cu mm.
- *Relation with pregnancy and labor*—in pregnancy and full term, the count is higher being highest during labor, i. e. 17,000 cells/mm³. There is also an increase during menstruation.

Types of WBC

- *Granular leukocytes or granulocytes*—it consist of neutrophils—62-70%, eosinophils—1-4% and basophils 0-1%.
- *Agranular leukocytes or agranulocytes*—it consist of lymphocytes (25 to 30%)—they are of two types, i.e. small lymphocytes and large lymphocytes and monocytes—5 to 10%.

Functions of WBC

- *Phagocytosis*—the neutrophils and the monocytes engulf and digest foreign bodies and bacteria.
- *Defensive mechanisms*—leukocytes play an important role in the defensive mechanism of the body by manufacturing fraction of serum globulin and formation of fibroblasts.
- *Trephones*—leukocytes manufacture trephones which have great influence on nutrition, growth and repair of tissue.
- *Heparin*—secretion of heparin which prevents intravascular clotting.
- *Anti-histaminic function*—it defends against allergic reactions.
- *Formation of fibroblasts*—lymphocytes may be converted into fibroblasts in areas of inflammation.

Platelets or Thrombocytes

They are non-nucleated, round or oval bi-convex discs having an average size of about 2.5 microns and are covered by a unit membrane. The average number of platelets in a

normal adult varies from 2,50,000 to 4,50,000 per cu mm. Vigorous muscular exercise, high altitude and hypoxia increase the platelet count.

Functions of Platelet

- *Hemostasis and blood clotting*—when there is bleeding, platelets disintegrate and liberate thromboplastin and 5-hydroxytryptamine. Thromboplastin activates prothrombin to form thrombin whereas 5-hydroxytryptamine has a vasoconstrictor effect and thus help in hemostasis.
- *Repair*—In circulation, platelets adhere to the damaged endothelial lining of the capillaries and thus bring about speedy repair.
- *Hasten clot retraction*—speed of clot retraction is directly proportional to the number of platelets present.

Blood Coagulation

When blood is shed, it loses its fluidity in few minutes and sets into semisolid jelly; this process is called as coagulation or clotting of blood.

Clotting Factors

- *Factor I or fibrinogen*—during clotting, fibrinogen is converted to fibrin to form clot.
- *Factor II or prothrombin*—during clotting, prothrombin is converted to thrombin by thromboplastin.
- *Factor III or thromboplastin*—it is essential for conversion of prothrombin into thrombin.
- *Factor IV calcium*—it is essential for the formation of both, intrinsic and extrinsic thromboplastin and also in the conversion of prothrombin to thrombin.
- *Factor V or labile factor or accelerator globulin or proaccelerin*—it is necessary for complete conversion of prothrombin into thrombin by extrinsic or intrinsic thromboplastin.
- *Factor VI or accelerin*—it is hypothetical activation product of proaccelerin.
- *Factor VII or stable factor*—factor X is converted to activated factor X by tissue thromboplastin released following tissue trauma, in the presence of factor VII for extrinsic pathway.
- *Factor VIII or anti-hemophilic factor or anti-hemophilic globulin or platelet cofactor*—it helps in the formation of intrinsic thromboplastin and prothrombin conversion.
- *Factor IX or Christmas factor or plasma thromboplastin component or platelet cofactor II*—it is necessary for intrinsic thromboplastin formation.
- *Factor X or Stuart factor*—it has properties similar to factor VIII.

- *Factor XI or plasma thromboplastin antecedent*—it is activated by active Hageman factor and ultimately leads to the formation of thrombin.
- *Factor XII or Hageman or surface factor*—it activates factor XI.
- *Factor XIII or fibrin stabilizing factor*—it, along with calcium ions convert soft fibrin clot into a solid fibrous one.

Mechanism of Blood Coagulation (Figs 37-1A to C)

The general mechanism takes place in three essential steps:

- *Thromboplastin release*—there is release of thromboplastin due to disintegration of platelets, when they come in contact with rough water, wettable surface and is also released from the damaged tissue.
- *Conversion of prothrombin into thrombin*—prothrombin activator catalyzes the conversion of prothrombin into thrombin.
- *Formation of clot*—thromboplastin acts as an enzyme for converting fibrinogen into fibrin threads that enmesh the blood cells and plasma to form the clot itself.

Blood Group

Human beings may be put into four group according to the nature of the agglutinogens possessed by their RBC. These are called as A, B, AB and O (if no agglutinin is present).

There are different types of Rh antigens each of which is called as Rhesus (Rh) factor. These types are designated as C, D, E. If D antigen is prominent, the person is said to be Rh positive.

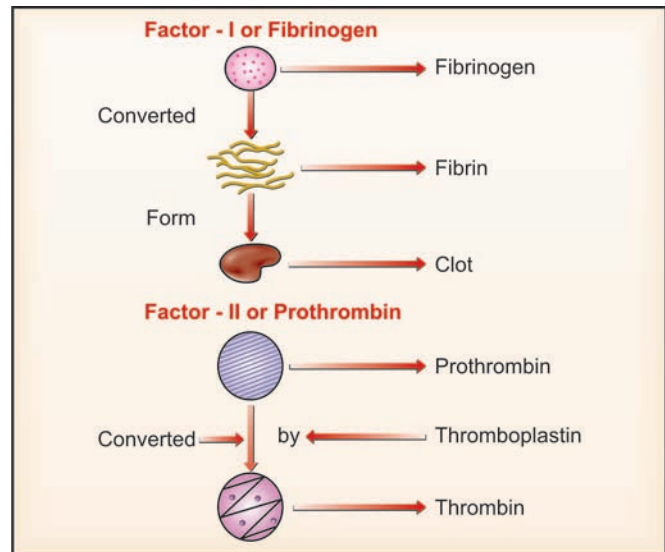


Fig. 37-1A

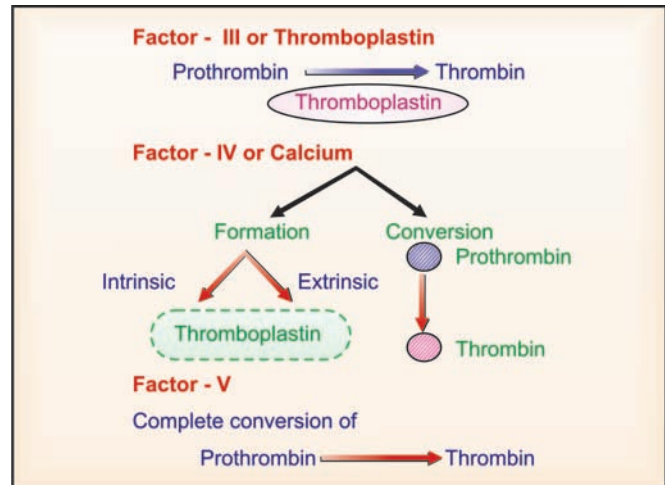


Fig. 37-1B

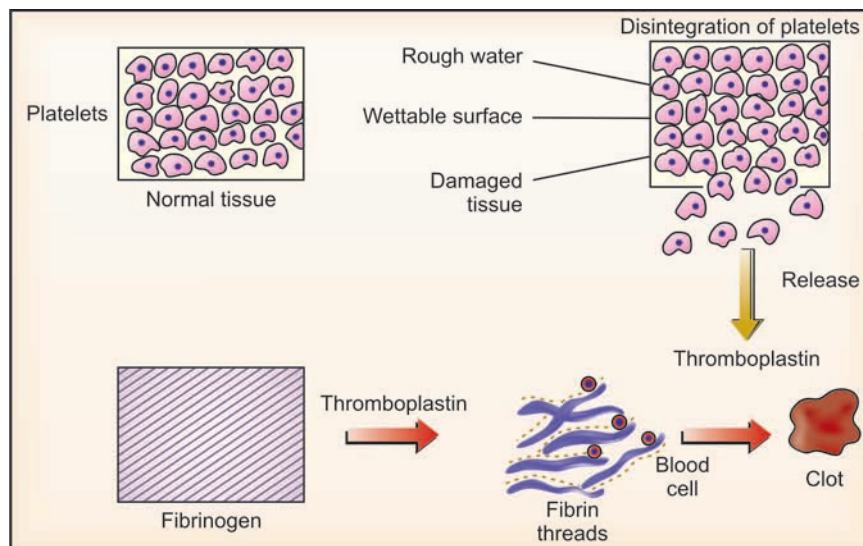


Fig. 37-1C

Figs 37-1A to C: A diagrammatic representation of mechanism of blood coagulation.

A person who has no D but instead has only C and E antigen is said to be Rh negative.

Diseases of Red Blood Cells

Anemia

It is an abnormal reduction in the number of circulating red blood cells, the quantity of hemoglobin and the volume of packed red cells in a given unit of blood.

Symptoms in a patient with anemia depend on following five factors:

- *Oxygen carrying capacity*—the reduction in the oxygen carrying capacity of the blood.
- *Total blood volume*—the degree of changes in total blood volume.
- *Rate*—the rate at which the above two have developed.
- *Capacity of cardiovascular and respiratory system*—the capacity of the cardiovascular and respiratory system to compensate for anemia.

Classification (Tables 37-1 to 37-3)

Anemia can be classified on the basis of etiology and morphology. Some of important anemias are described below.

Posthemorrhagic Anemia

The anemia caused by blood loss may occur in variety of conditions causing bleeding.

Types

- *Acute*—when blood loss occurs in large amounts in a short period of time, anemia may develop even though iron stores remain adequate. It is called as acute post-hemorrhagic anemia.
- *Chronic*—this develops when there is chronic blood loss in small amount over prolonged period of time and is due to depletion of iron in body. Other findings are the same as that of iron deficiency anemia.

Clinical Features

- *Factors affecting the clinical features*—the manifestations of hemorrhagic anemia depend on the rate and magnitude of bleeding; the time elapsed since it took place and the site—whether it is external or internal.
- *Blood loss (500 to 1000 ml)*—when the blood loss is about 500 to 1000 ml, most of the patients do not present any symptoms, but few may present with weakness and sweating.
- *Blood loss (1000 to 1500 ml)*—with rapid loss of 1000 to 1500 ml, a previously healthy individual may experience lightheadedness and hypotension.

Table 37-1: Classification of anemia (according to etiology)

Loss of blood

- Acute posthemorrhagic anemia
- Chronic posthemorrhagic anemia

Excessive destruction of RBCs

- *Extracorporeal causes*
 - Antibodies
 - Infections like malaria
 - Splenic sequestration and destruction
 - Associated diseases like lymphomas
 - Drugs, chemical and physical agents
 - Trauma to RBC
- *Intracorporeal hemolytic diseases*
 - *Hereditary*
 - Disorders of glycolysis
 - Faulty synthesis or maintenance of reduced glutathione
 - Qualitative or quantitative abnormalities in the synthesis of globulin
 - Abnormalities in RBC membrane
 - Erythropoietic porphyria
 - *Acquired*
 - Paroxysmal nocturnal hemoglobinuria
 - Lead poisoning

Impaired blood production resulting from deficiency of substances essential for erythropoiesis

- Iron deficiency
- Deficiency of vitamin B₁₂, folic acid (pernicious anemia and megaloblastic anemia)
- Pyridoxine responsive anemia
- Protein deficiency
- Ascorbic acid deficiency

Inadequate production of mature erythrocytes

- *Deficiency of erythroblast*
 - *Aplastic anemia* (atrophy of bone marrow)
 - Chemical or physical agents
 - Hereditary
 - Idiopathic
 - Pure red cell aplasia (isolated erythroblastopenia)
 - Thymoma
 - Chemical agents
 - Antibodies
- *Infiltration of bone marrow*
 - Leukemia
 - Multiple myeloma
 - Carcinoma
 - Sarcoma
 - Myelofibrosis
- *Endocrine abnormalities*
 - Myxedema
 - Addison's disease (adrenal insufficiency)
 - Pituitary insufficiency
 - Hyperthyroidism
- *Chronic renal failure*
- *Chronic inflammatory disease*
 - Infectious
 - Noninfectious including granulomatous and collagen disease
- *Cirrhosis of liver*

Table 37-2: Morphologic classification of anemia

Type of anemia	Description	Most common causes
Macrocytic	Increased MCV, MCH and normal MCH conc.	Lack of erythrocyte maturing factors
Normocytic	Reduction only in RBC membrane; normal MCV, normal MCH and normal MCH conc.	Hemorrhage, hemolysis lack of blood formation, dilution of blood with fluid
Simple microcytic	Reduced MCV, MCH, MCH conc.	Associated with infections and inflammatory diseases
Hypochromic microcytic	Reduced MCV, MCH, MCH conc.	Iron deficiency

Table 37-3: Third classification

Anemias with disturbed iron metabolism
• Iron deficiency anemia
• Sideroblastic anemia
Megaloblastic anemia
• Pernicious anemia (vitamin B ₁₂ deficiency)
• Folic acid deficiency
Anemia associated with chronic disorders
• Anemia of chronic infection
• Anemia of inflammatory connective tissue disorders
• Anemia associated with malignancy
• Secondary to chronic bleeding
• Myelophthisic anemia
• Anemia of uremia
• Anemia of endocrine failure
• Anemia of liver disease
• Hemolytic anemia
• Extrinsic causes
• Splenomegaly
• Red cell antibodies
• Trauma in the circulation
• Direct toxic effects
• Membrane abnormalities
• Spur cell anemia
• Paroxysmal nocturnal hemoglobinemia
• Hereditary spherocytosis
• Hereditary elliptocytosis
• Disorders of interior of the red cell
• Defects in the Embden-Meyerhof pathway
• Defects in the hexose monophosphate shunt
Disorders of hemoglobin
• Sickle cell anemia
• Thalassemia

- **Blood loss (1500 to 2000 ml)**—when 1500 to 2000 ml of blood is lost, symptoms like sweating, thirst, shortness of breath, clouding or loss of consciousness are seen. The pulse becomes rapid and low in volume, skin become cold and clammy and there is a fall in blood pressure.
- **Blood loss > 2000 ml**—when rapid loss of blood exceeds 2000 to 2500 ml, severe state of shock is reached.

Diagnosis

- **Clinical diagnosis**—history of blood loss with features of anemia may diagnose this condition.
- **Laboratory diagnosis**—plasma volume and red cell mass are reduced in proportional amount. Anemia is

normocytic and normochromic. Erythropoietin secretion is stimulated which in turn gives rise to hyperplasia of marrow erythroid elements within 3 to 5 days. There is increased in reticulocyte number. Neutrophilic leukocytosis often follows hemorrhage and maximum after 2-5 hours.

Management

- **Restoration of blood volume**—blood transfusion should be given to restore blood volume.
- **Other therapy**—intravenous infusion of saline, dextrin, albumin or plasma.

Iron Deficiency Anemia

Iron is essential for synthesis of 'heme' portion of hemoglobin. Iron deficiency anemia is caused by imbalance between iron intake and loss or inadequate utilization.

Causes

- **Inadequate intake**—inadequate intake of iron in the diet may lead to deficiency.
- **Malabsorption**—malabsorption of iron due to hypochlorhydria and diarrhea may lead to deficiency state.
- **Increase requirement**—increased requirement of iron in a growing child and in pregnancy.
- **Increases loss**—increased loss of iron due to injury, recurrent epistaxis and peptic ulcer. Chronic blood loss such as menstrual and menopausal bleeding parturition.
- **Gastrotomy**—subtotal or complete gastrotomy may cause iron deficiency anemia.

Stages of Iron Deficiency

- **Prelatent iron deficiency**—in this, body storage of iron are depleted but the circulating hemoglobin and serum iron remain within the normal limit.
- **Latent iron deficiency**—in it, body storage of iron are exhausted and serum iron is reduced. The hemoglobin concentration remains unaffected.

- *Early anemia*—the third stage is iron deficiency anemia in which the hemoglobin concentration is reduced in peripheral blood. The serum iron is low.
- *Marked anemia*—in this, hemoglobin concentration is below 10 g per 100 ml of blood. Red blood cells show morphological changes in this stage.

Clinical Features

- *Age and sex*—it occurs chiefly in women in the 4th and 5th decades of life.
- *Symptoms*—the patient experiences tiredness, headache, paresthesia and lack of concentration.
- *Koilonychia*—nails become brittle, flattened and often show spoon shape (koilonychia).
- *Neuropathy*—there may be tingling and pins and needle sensation in the extremities.
- *Dysphagia*—some patients develop pharyngeal mucosal thickening and mucosal web formation, giving rise to dysphagia.
- *Gastrointestinal symptoms*—liver and spleen may be palpable. There may be gastrointestinal bleeding and menorrhagia there by setting a vicious circle.
- *Knuckle pigmentation*—patient may get knuckle pigmentation (pigmentation seen on dorsum of hand at metacarpophalangeal joint (**Fig. 37-2**).



Fig. 37-2: Knuckle pigmentation seen on dorsum of hand at metacarpophalangeal joint

- *Sideropenic anemia*—in certain pathological conditions, iron gets accumulated in mitochondria and appears as a ring of granules round the nucleus. These are called as ring sideroblasts and are the characteristic cells of sideropenic anemia which is either hereditary or acquired. Acquired type can be associated with rheumatoid arthritis, malignant diseases, pernicious anemia and myxedema. Clinical features are same as those of iron deficiency anemia.

Oral Manifestations

- *Pallor of mucosa*—in iron deficiency there is pallor of oral mucosa and gingiva. The normal pink color is lost due to lack of oxygenated blood in the capillary bed in lamina propria and is associated with lowered levels of hemoglobin.
- *Atrophy of oral mucosa*—the generalized atrophy of oral mucosa both in tongue and buccal mucosa occurs.
- *Tongue changes*—there is redness, soreness or burning of tongue. The filiform papillae over the anterior two thirds of tongue are the first to undergo atrophy (**Fig. 37-3**). In severe cases, fungiform papillae are also affected leaving the tongue completely smooth and waxy or glistening in appearance (**Fig. 37-4**).



Fig. 37-3: Atrophy of tongue papillae seen in iron deficiency anemia (Courtesy Dr Milind Chandurkar).

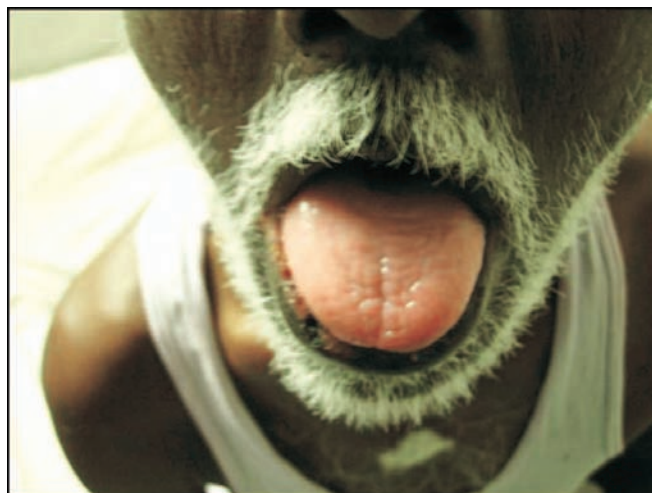


Fig. 37-4: Pallor of tongue with bald appearance seen in iron deficiency anemia (Courtesy Dr Milind Chandurkar).

- *Angular cheilitis*—there is cracking and fissuring at the corner of mouth.
- *Ulceration*—there is softening of epithelium which leads to linear ulceration of the skin, extending up to and beyond the mucocutaneous junction. There may be pain or bleeding from ulcerated tissues. Recurrent aphthous ulceration and candidal lesions can also occur in iron deficiency anemia.
- *Slow healing*—patient may show slow healing after oral surgical procedures.
- *Gingival enlargement*—in some cases, there may be gingival enlargement.

Diagnosis

- *Clinical diagnosis*—koilonychia, neuropathy, angular cheilitis and depapillation of tongue may yield in diagnosis.
- *Laboratory diagnosis*—the anemia is microcytic and hypochromic and the peripheral smear show abnormal forms of RBCs. There is reduced hemoglobin level, as low as 4g/100ml. There is normal or slightly reduced RBC count. MCV, MCH and MCHC are all reduced.

Management

- *Iron supplement*—almost all patients can be treated by oral supplements of iron by giving ferrous fumarate or ferrous sulphate. It is given in dose of 300 mg three to four times a day for a period of 6 months. The parenteral route of administration is suitable for few patients who are unable to take iron by mouth or who are unable to absorb iron by mouth. The recommended single dose of iron sorbitol is 1.5 mg of iron per kg of body weight, given daily.

Plummer-Vinson Syndrome

It is also called as *Paterson-Brown-Kelly syndrome*. It is characterized by dysphagia, iron deficiency anemia, dystrophy of nails (koilonychia) (**Fig. 37-5**) and glossitis.

Clinical Features

- *Age*—it is exclusively found in middle aged women.
- *Appearance*—patient of this syndrome have got characteristic asthenic appearance.
- *Dysphagia*—it occurs due to the formation of webs in esophagus.
- *Angular cheilitis*—vermillion borders of the lip are very thin and there is often angular cheilosis.
- *Symptoms*—patients complaint of ‘spasm in throat’ or food sticking in throat. There is also complaining of sore mouth and inability to retain dentures.
- *Tongue*—A smooth, red, occasionally enlarged and often sore tongue with fissuring is occurs.

- *Signs*—the width of mouth is narrowed and the oral mucosa is pale and painful. There is also dry mouth and spoon shaped nails. There are atrophic changes in mucosa of mouth, pharynx, upper esophagus and vulva.
- *Koilonychia*—this is also present in case of Plummer-Vinson syndrome.



Fig. 37-5: Koilonychia (spoon shaped nail) seen in Plummer-Vinson syndrome (Courtesy Dr Milind Chandurkar).

Diagnosis

- *Clinical diagnosis*—dysphagia, glossitis, with features of iron deficiency anemia will diagnose Plummer-Vinson syndrome.
- *Laboratory diagnosis*—same features as that of iron deficiency anemia. Biopsy will show atrophic epithelium and atrophy of lamina propria and muscles.

Management

- *Correction of anemia*—management is directed management of iron deficiency anemia
- *Esophageal dilation*—esophageal dilation is required to correct dysphagia.

Erythroblastosis Fetalis

It is type of isoimmune hemolytic anemia. It occurs due to isoimmune antibodies. It is also called as ‘*hereditary disease of new born*’ (HDN). Congenital hemolytic anemia due to Rh incompatibility results from destruction of fetal blood brought about by a reaction between maternal and fetal blood factors. The Rh factor, named after *Rhesus monkey*, was discovered by Landsteiner and Wiener in 1940 as a factor in human RBC which reacts with rabbit antiserum produced by administration of red blood cells from Rhesus monkey.

Pathogenesis

- *Inheritance*—it occurs due to inheritance. Blood factor from the father acts as a foreign antigen to mother fetus.
- *Formation of antibodies*—there is transplacental transfer of father antigen from fetus to mother. This will result in immunization of mother with formation of maternal antibodies against fetal antigens. If the father is Rh positive and the mother is Rh negative, fetus inherits Rh-positive antigens, which may act as antigen to the mother and immunize her with resultant antibody formation.
- *Destruction of fetal red blood cells*—some of these antibodies then cross into fetal circulation and cause the destruction of fetal red cells.
- *Complicating factors*—the problem is complicated by Rh antigens which are termed as C, D and E. Out of this; D antigen is the strongest and is responsible for the clinical manifestations of erythroblastosis fetalis.

Clinical Features

- *Stillborn*—some infants are stillborn.
- *Symptoms*—children having pallor, jaundice, compensatory erythropoiesis (both medullary and extra-medullary) and edema resulting in fetal hydrops.
- *Kernicterus*—it may manifest itself by apathy and poor feeding and later by mental retardation, irritability and cranial nerve palsies.

Oral Manifestations

- *Teeth*—it may manifest in teeth by the deposition of blood pigment in enamel and dentin of developing teeth giving them a *green, brown or blue hue*. It occurs in those portions of the teeth which are being laid down during the time when icterus was at its height.
- *Enamel hypoplasia*—enamel hypoplasia is also reported occurring in some cases of erythroblastosis fetalis. It usually involves incisal edges of the anterior teeth and middle portion of the deciduous cuspid and first molar crown.
- *Rh hump*—there is ring like defect called as 'Rh hump'.

Radiographic Features

- *Radiodensity*—there is presence of transverse lines of increased and decreased radiolucency, occurring at the ends of some long bones in some areas.
- *Cranial vault*—there is also increased thickness of cranial bone which includes maxilla and mandible. The bones of vault are thickened as a result of increased thickness of diploe.
- *Radiating spicule*—radiating spicule may also be present.

Diagnosis

- *Clinical diagnosis*—children having kernicterus, Rh hump and enamel hypoplasia will diagnose the case.
- *Laboratory diagnosis*—the red blood count at birth may vary from less than the 1,000,000 cells per cubic millimeter to near a normal level. The icterus index is very high and may reach a level of 100 units. The peripheral smear shows large number of immature RBCs. There is also evidence of hemolysis.

Management

- *Antenatal*—maternal sensitization is prevented by the administration of 300 mg of Rh antiglobulin after amniocentesis has been done. Management of affected fetus can be done by intensive plasmapheresis of mother during pregnancy in order to remove maternal antibodies or by intravenous transfusion of blood into the fetus to prevent fetal anemia.
- *Post natal*—in infants, exhibiting only anemia, fresh packed RBCs are given to correct anemia.
- *Splenectomy*—it results in striking and permanent improvement, both in anemia and symptoms. After splenectomy, daily penicillin V 250 mg 12 hourly is given to avoid infections.
- *Blood transfusion*—severe hemolytic crisis needs blood transfusion. Folic acid should be given 5 mg orally to support increased erythropoiesis.
- *Vaccination*—the patient should be vaccinated against pneumococci and hemophilus influenzae.

Sickle Cell Anemia

It is chronic hemolytic blood disorder characterized by abnormal hemoglobin (deoxygenated hemoglobin) which under low oxygen tension, results in sickling of cell. It is severe genetic disorders of hemoglobin synthesis. It is autosomal dominant. It was first described by Herrick in 1910.

Pathogenesis

- *Substitution of amino acid*—substitution of amino acid glutamine on position 6 present in the chain of the hemoglobin-A, by valine.
- *Formation of hemoglobin S*—this will give rise to an abnormal hemoglobin, i.e. hemoglobin S.
- *Formation of tactoids*—when Hbs are deoxygenated, it forms structures known as 'tactoids'.
- *Sickle shaped cells*—tactoids will distort the RBC membrane and produce characteristic sickle shaped cell which are destroyed by RE cells.
- *Thrombosis and infarction*—sickle cells increase blood viscosity and tend to reduce blood flow leading to thrombosis and tissue infarction.

- *Hemolysis*—in addition, these cells are phagocyte in large number by mononuclear-phagocyte system which reduce their lifespan and give rise with hemolysis.
- *Folic acid deficiency*—patients may develop severe folic acid deficiency due to increased erythropoiesis.

Types

- *Sickle cell disease*—in homozygous individuals, whole of hemoglobin-A is replaced by hemoglobin-S and this is known as 'sickle cell disease'.
- *Sickle cell trait*—heterozygous individuals, only 50% of HbA is replaced by HbS and this is known as 'sickle cell trait'.

Clinical Features

- *Sex and age*—it is common in females and mostly the clinical symptoms become evident before the age of 30 years.
- *Incubation period*—clinical manifestations begin only after several months as fetal hemoglobin protects against sickling phenomenon.
- *Precipitating factors*—include dehydration, chills and infection but sometime, the attack occurs spontaneously.
- *Symptoms*—there is fatigue, weakness and shortness of breath. Patient may complaint of severe abdominal pain, muscle and joint pain, at high temperature which may result in circulatory collapse. There is painless hematuria.
- *Sign*—pallor on palms (**Fig. 37-6**), there is enlargement of heart and murmur is found in most of the patients. There is increased susceptibility to infection. Most of persons expire before the age of 40 years. There is also presence of leg ulcer and gallstones. When associated with folate deficiency, there may be growth retardation and also delayed puberty.



Fig. 37-6: Pallor is seen on hand of patient with sickle cell anemia. To compare between pallor with normal hand, another hand is kept for the comparison (Courtesy Dr Milind Chandurkar).

- *Skull changes*—hyperplasia of marrow in first year of life expands the marrow cavity producing bossing of the skull, prominent malar bones and protuberant teeth.
- *Sickle cell crisis*—in this, sickling becomes severe after long quit spell of hemolytic latency. Hypoxia, infection, hypothermia or dehydration can precipitate the crisis. Patient experience severe pain due to ischemia. Each episode occasionally punctuated by exacerbations called as sickle cell crisis.
- *Acute chest syndrome*—pulmonary involvement is called as acute chest syndrome. This is serious complication of sickle cell crisis.
- *Infarction*—infarction can only occur in bone and spleen but, other tissues may also be involved. In infants, fingers and toes are commonly affected.
- *Thrombosis*—thrombosis of vessels in brain cause severe neurologic disorders like stroke, convulsion, coma, downiness as well as speech visual and hearing disturbances. Occlusion of smaller vessels results in headache and cranial nerve neuropathy including palsy and paresthesia.

Oral Manifestations

- *Oral mucosa*—the oral mucosa will show pallor and jaundice.
- *Teeth*—there may be delayed eruption and hypoplasia of the dentition, secondary to their general development.
- *Osteomyelitis*—patient is more prone to develop osteomyelitis. This may be due to hypovascularity of the bone marrow secondary to thrombosis.
- *Nerves*—patient may present with paresthesia of mental nerve which may be secondary to occlusion involving the nerves and blood supply.
- *Jaw bones*—mongoloid facies with high cheek bones and bimaxillary prognathism. It is due to marrow hyperplasia resulting in an increase in hard palate length and palate alveolar ridge angle.

Radiological Findings

- *Osteoporosis*—because of chronic increased erythropoietic activity and marrow hyperplasia, there is loss of trabeculation of the jaw bone resulting in a mild to severe generalized osteoporosis and appearance of large irregular marrow spaces.
- *Ground glass appearance*—the increased radiolucency of the jaws which is seen to have a ground glass appearance which is attributed to diminution of fine trabeculae as there is replacement of marrow by compensatory hyperplasia.
- *Inferior border of the mandible*—thinning of inferior border of mandible of alveolar cortex.

- *Lamina dura*—lamina dura appears more prominent than normal, particularly against the background of the increased radiolucency of the bone.
- *Step ladder effect*—the trabeculae in between the roots of teeth may appear as horizontal rows creating 'step ladder' like effect.
- *Maxilla and teeth*—enlargement of maxillae with protrusion and separation of upper anterior teeth.
- *Hair on end appearance*—skull radiographs show an unusual appearance, with perpendicular trabeculation radiating outward from the inner table producing a 'hair on end' pattern. There is thickening of the frontal and parietal bones. The outer table of bone may appear to be absent and diploes are thickened.

Diagnosis

- *Clinical diagnosis*—severe abdominal pain, mongoloid facies with prominent cheek bone and bimaxillary prognathism.
- *Radiological diagnosis*—hair on end appearance, step ladder effect and osteoporosis of jaw bone will diagnose sickle cell anemia.
- *Laboratory diagnosis*—a smear of peripheral blood usually shows typical sickle shaped red blood cells and hemoglobin level is decreased. The RBC count may reach level of 10 lac cells or less per cu mm with Hb levels from 5 to 12 gm/dl. The sickle hemoglobin molecule undergoes gelation or crystallization, when deoxygenated and this typically distorts the erythrocyte, producing the sickle shape. This sickle shaped cells then 'logjam' and produce stasis within the microvasculature. Damage to erythrocyte membrane also occurs in sickle cells and leads to their fragmentation and intravascular hemolysis.

Management

- *Prevention*—prevention of episode by avoiding the precipitating factors is an important aspect of treatment. Patients should avoid becoming chilled, dehydrated or exposed to hypoxia (high altitude).
- *Folic acid*—regular folic acid supplement (5 mg/daily) is given to support the greatly increased erythropoietin.
- *Antibiotics*—prophylactic antibiotics are given to prevent infections. Young patient with hyposplenism should be given phenoxymethyl penicillin.
- *Analgesics*—pain caused by tissue infarction is extremely severe but analgesics should be avoided or if at all necessary, no addictive analgesics like aspirin, paracetamol should be given because the episodes are recurrent.
- *Hydroxyurea*—this is indicated in adult patients in severe cases. The drug increases fetal form of hemoglobin which inhibits the polymerization of hemoglobin S and reduce

adherence of erythrocytes to vessels walls. It has got potential side effect and should be used cautiously.

- *Blood transfusion*—blood transfusion should be given during crisis only. Routinely, it should be avoided, because it increases the viscosity of blood.
- *Genetic counseling*—it should be done for patient who has sickle cell trait.

Dental Considerations

- *Shorten time for dental procedure*—do not prolong or carry out extensive dental procedures involving the soft tissues unless absolutely necessary. Keep the dentition as healthy as possible because there are always chances that any infection might precipitate an aplastic crisis.
- *Anesthesia*—avoid using general anesthesia or when used, it is imperative to avoid episodes of hypoxia because of the cerebral or myocardial thrombosis which might result.

Thalassemia

It is also called as 'Cooley's anemia', 'Mediterranean anemia' and 'erythroblastic anemia'. As majority of the cases of thalassemia are seen in the region of Mediterranean Sea, the name thalassemia is given, thalassa stands for sea. It is autosomal dominant.

It is an inherited impairment of hemoglobin synthesis in which there is partial or complete failure to synthesize a specific type of globin chain. Beta thalassemia is more commonly occur as compared to alpha thalassemia.

Pathogenesis

- *Absence of alpha or beta chains*—hemoglobin molecule composed of two alpha and two beta chains. Either alpha or beta globulin genes may be affected.
- *Reduces hemoglobin*—if one of chains is not made adequately, the resultant red blood cells have reduced hemoglobin. This will result in shortened lifespan of RBCs.
- *Alteration in structure of cells*—the excess globin chains accumulate within the erythrocytes, which results in alteration in structure of cells.
- *Hemolysis*—these abnormal erythrocytes got destructed in spleen (hemolysis) and patient suffers from hypochromic and microcytic anemia.
- *Bone marrow hyperplasia*—this occur as rare of hematopoiesis is increased to maintain adequate oxygenation.

Types

- *Alpha thalassemia*—there is reduction or absence of α chain synthesis. Alpha chains of hemoglobin are required not only for HbA but also for HbF, which is the

main hemoglobin type in fetal life. Therefore, major type of alpha thalassemia is incompatible with life and results in hydrops fetalis and intrauterine death of fetus.

- *Beta thalassemia*—there is reduction or absence of beta chains. Hemolysis is not primarily due to lack of β -globin chains but it is because of the free alpha chains which form insoluble aggregates that precipitate within the RBCs and cause damage to the cell membranes.
- *Thalassemia major or homozygous β -thalassemia*—occurs when the patient is homozygous. It is also called as Cooley's anemia. In this case two defective gene is affected.
 - *Hemoglobin H disease*—it is very mild form of the disease in which the patient may live relatively normal life.
 - *Hemoglobin Bart disease*—in which infants are stillborn or die shortly after birth.
- *Thalassemia intermedia*—it is group of disorders characterized by clinical manifestations between major and minor.
- *Thalassemia minor or thalassemia trait*—occurs when the patient heterozygous. In this, only one defective gene is affected.

Clinical Features

- *Age*—occurs between the ages of 6 to 24 month and after the age of 6-8 months.
- *Symptoms*—the patient first presents with pallor of skin, fever, chills, malaise, generalized weakness, prominent cheek bone and mild hepatosplenomegaly.
- *Mongoloid appearance*—bone marrow hyperplasia in early life may produce frontal head bossing and there may be marked overdevelopment of malar bone which is associated with a short nose having a depressed bridge giving the appearance of mongoloid.
- *Organs failure*—deposition of iron in various organs (due to multiple transfusions) leads to signs and symptoms of organ failure.
- *Prognosis*—most patients die in childhood due to anemia and cardiac failure.
- *Alpha thalassemia*—the clinical findings are severe alveolar bone loss, pronounced spacing of maxillary anterior teeth and mongoloid appearance.

Oral Manifestations

- *Jaw bone*—there is excessive overgrowth of maxilla causing excessive lacrimation and nasal stuffiness.
- *Oral mucosa*—the oral mucosa is pale and has a lemon yellow tint because of chronic jaundice. The color is best seen at the termination of hard palate and in the floor of mouth.

- *Rodent facies*—there is marked over development of maxilla associated with hyperplasia of alveolar process, which results in anterior open bite and prominent cheek bone producing characteristic 'rodent facies'.
- *Chipmunk facies*—there is also saddle nose, prominent malar bone and pneumatization of maxillary sinus. As a result of these skeletal changes, the upper lip is retracted giving the child a 'chip munk' facies.
- *Teeth*—the maxillary teeth are protrudes with spacing between them with a short upper lip due to a lag between the growth of the maxilla and the growth of upper lip. Due to high concentration of iron, discoloration of dentin and enamel may be evident.
- *Poor healing*—there is poor healing after dental treatment.

Radiographic Features

- *Teeth*—the root of mandibular 1st molar and central incisors may be spike shaped and short. The lamina dura is thin and the roots of the teeth may be short. Circular radiolucency may be seen in the lower anterior region. There is thinning of the crypt of developing teeth.
- *Alveolar bone*—the alveolar bone shows large bone marrow spaces, osteoporosis and blurring of trabeculae. The marrow spaces are large and the trabeculae are large and course.
- *Compensatory lamellar striation*—some areas may show prominent trabeculae which have been referred to as 'compensatory lamellar striation'.
- *Long bones*—in the long bone there is generalized radiolucency, increased width of shaft, and narrowing of cortices due to the greater amount of marrow substance. There is also pathological fracture.
- *Skull*—skull is thickened as a result of increased width of the dipole space between the outer and inner tables of the vault. This occurs due to proliferation of the hemopoietic tissue. Generalized granular appearance in skull.
- *Antrum*—the antra is reduced in size by the encroachment of the bone formation, which is sometimes sufficient to cause complete effacement of the air sinuses.
- *Premaxilla*—premaxilla is prominent resulting in malocclusion.
- *Cortical borders*—there is generalized thinning of cortical borders.
- *Hair on end appearance*—the trabeculae joining the inner and outer table of the skull are readily arranged calcified spicules which appear as calcified hair extending between the inner and outer tables, which is called as 'hair on end' appearance (Fig. 37-7).

Diagnosis

- *Clinical diagnosis*—mongoloid appearance, chipmunk facies, and rodent facies with discoloration of dentin may give clue to diagnosis of thalassemia.

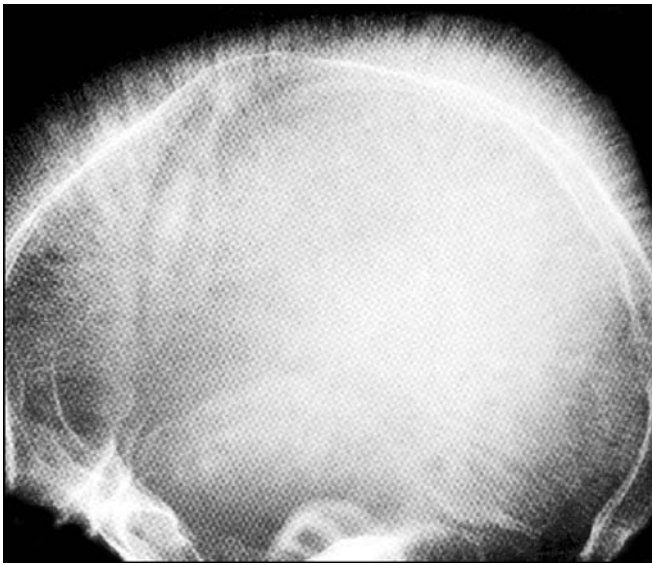


Fig. 37-7: Typical hair on end appearance seen in thalassemia patient (Courtesy Dr Ganju).

- **Radiological diagnosis**—hair on end appearance, compensatory lamellar striation with spike shaped mandibular molar.
- **Laboratory diagnosis**—the anemia is hypochromic and microcytic. The peripheral smear shows abnormal RBCs. Reticulocyte count is increased. Bone marrow shows increased erythropoietic activity. Serum bilirubin and fecal and urinary urobilinogen are elevated because of severe hemolysis. An elevated fetal hemoglobin is present. Biopsy shows typical safetypin cells in the circulation.

Management

- **Blood transfusion**—repeated blood transfusions are necessary. Repeated blood transfusion will lead to increased iron load in the tissue of the body. This will lead to a condition known as *hemochromatosis*.
- **Chelating agent**—iron overload can be reduced by regular use of chelating agents like *desferrioxamine* therapy.
- **Bone marrow transplantation**—allogenic bone marrow transplantation can be done. It is indicated in young individual, little organ damage and HLA matched donor.
- **Splenectomy**—splenectomy is indicated when there is an increase in the severity of anemia.
- **Folic acid**—the patient may need supplementary folic acid because of increased requirement owing to increased erythropoiesis.
- **Surgical correction**—it is indicated in patient who developed an abnormal facial appearance.

Erythropoietic Porphyria

It is a group of inherited or acquired disorders characterized by excessive production, accumulation and excretion of

porphyrins and their precursors. It is a metabolic defect within the maturing erythrocytes resulting in excessive production of uroporphyrinogen-I.

Clinical Features

- **Age**—it occurs soon after birth.
- **Symptoms**—the urine is burgundy red in color or turns red on exposure to light.
- **Signs**—photosensitivity of exposed parts of body leads to formation of blister and scars.
- **Anemia**—there is also hemolytic anemia and splenomegaly.

Oral Manifestations

- **Erythrodontia**—there is deposition of porphyrin in dentin, to a lesser extent in the enamel which imparts red or brown color to deciduous and permanent teeth (erythrodontia). The staining by uroporphyrin can be confirmed by ultraviolet light which will produce fluorescence.
- **Signs**—bullous erosions of oral mucosa can be seen. In advanced cases, pigmented atrophic scars on lip are seen resemble the keloid structures. The patient is hence, unable to close his mouth.

Diagnosis

- **Clinical diagnosis**—burgundy red color urine which turns red upon exposure to light with erythrodontia will diagnose the disease.
- **Laboratory diagnosis**—about 50% normoblasts, reticulocyte and 50% RBCs exhibit intense red fluorescence under fluorescence microscope.

Management

- **Splenectomy**—in severe cases of hemolysis, splenectomy may be carried out.

Megaloblastic Anemia

Megaloblastic anemia occurs due to deficiency of vitamin B₁₂ or folate or both, resulting in disordered cell proliferation leading to megaloblastic anemia. All megaloblastic anemias have erythroid precursors known as megaloblasts.

Causes

- **Folate deficiency**—the condition is prevalent in women during pregnancy and lactation. It may occur due to low availability of folate in diets, prolonged cooking of food, malabsorption syndromes such as tropical sprue and malaria.

- **Vitamin B₁₂ deficiency**—removal of vitamin B₁₂ from the gut either by bacterial proliferation or by parasites. It may occur due to chronic dietary deficiency of the vitamin.

Clinical Features

- **Symptoms**—there are symptoms of anemia like weakness, loss of appetite and palpitations. Patient may get periodic diarrhea.
- **Skin**—in severe cases, skin may show faint lemon yellow tint and spleen may be palpable.
- **Neuropathy**—many cases show paresthesia of finger and toes and dementia may also be seen.

Oral Manifestations

- **Symptoms**—burning sensation in tongue, hypersensitivity, paresthesia and later dryness of the mouth.
- **Hunter's glossitis**—atrophy of filiform and fungiform papillae leads to completely atrophic smooth fiery red surface of tongue (Hunter's glossitis) (**Fig. 37-8**).
- **Angular cheilitis**—angular cheilitis and dysphagia due to pharyngitis and esophagitis.
- **Oral mucosa**—yellowish brown pigmentation of oral mucosa may occur due to increased circulating bile pigments.



Fig. 37-8: Hunter glossitis showing fiery red appearance in megaloblastic anemia.

Diagnosis

- **Clinical diagnosis**—symptoms of anemia with Hunter glossitis with dysphagia may give clue to diagnosis.
- **Laboratory diagnosis**—the peripheral smear shows a macrocytic blood picture with many abnormal forms of RBCs. There is associated leukopenia and sometimes, thrombocytopenia. Bone marrow shows presence of megaloblasts and erythroid hyperplasia. In advanced

cases, the red blood cell abnormalities are detected like polychromatophilic cells, stippled cells, nucleated cells, Howell-Jolly bodies and Cabot's ring.

- **Bone marrow finding**—there is increased number of immature red cells or megaloblasts with a few normoblast. Polymacocytes or large PMNs with large polylabeled nuclei are found. Megakaryocytes appear normal.

Management

- **Education**—general education on cooking practices and food habits is undertaken in tropical countries to prevent nutritional anemia.
- **Blood transfusion**—when hemoglobin level is less than 4 gm/dl blood, transfusion should be given.
- **Vitamin B₁₂**—oral administration of vitamin B₁₂ should be given.
- **Folic acid**—folic acid supplements should also be given.

Pernicious Anemia

It is also called as 'primary anemia', 'Addison's anemia' or 'Biermer's anemia'. The term pernicious anemia should be reserved for patients who have B₁₂ deficiency secondary to intrinsic factor deficiency.

Causes

- **Atrophy of gastric mucosa**—it occurs due to atrophy of gastric mucosa resulting in failure to secrete the still unidentified 'intrinsic factor'.
- **Autoimmune disorders**—it is suggested that it is autoimmune disorder, because autoantibodies to gastric parietal cells are often found in patients.

Clinical Features

- **Age**—it is rare before the age of 30 years and increase in frequency with advancing age. Males are more commonly affected than females.
- **Symptoms**—there is usually triad of symptom: generalized weakness, sore painful tongue, numbness and tingling of the extremities. Other features are fatigability, headache, dizziness, nausea, vomiting, diarrhea, loss of appetite, shortness of breath, loss of weight, pallor and abdominal pain.
- **Nervous system**—nervous system disorders are also present and manifested by sensory disturbances including the paresthetic sensation of the extremities, weakness, stiffness and difficulty in walking, general irritability, depression, drowsiness as well as incoordination on loss of vibratory sensation. There is also tingling sensation in the fingers and toes that eventually progress to numbness.
- **Gastrointestinal complaint**—Epigastric discomfort, constipation or diarrhea can be seen in these patients.

Oral Manifestations

- **Symptoms**—glossitis (**Fig. 37-9**) and patient complains of painful and burning lingual sensation which may be so annoying that the dentist is often consulted first. Sometimes, inflammation and burning involve the entire oral mucosa. There is disturbance in taste sensation with intolerance to dentures and occasional dryness of mouth.



Fig. 37-9: Glossitis in patient with pernicious anemia.

- **Beefy red tongue**—the tongue is generally inflamed often described as 'beefy red' in color, either entirely or in patches scattered over the dorsum and lateral border of tongue.
- **Hunter glossitis or Moeller glossitis**—there is gradual atrophy of the papillae of tongue that eventuates in a smooth and bald tongue which is often referred as *Hunter's glossitis* or *Moeller's glossitis* and is similar to the 'bald tongue of sandwich' seen in pellagra. The fiery red appearance of tongue may undergo remission but recurrent attacks are common.
- **Xerostomia**—patient may notice xerostomia. Xerostomia may lead to occurrence of lobulation on tongue.
- **Oral mucosa**—oral mucosa shows greenish yellow color (frequently observed on the skin) at the junction of hard and soft palate, when daylight is used for illumination.

Diagnosis

- **Clinical diagnosis**—triad of generalized weakness, sore and painful tongue and numbness and tingling of extremities are typical of pernicious anemia. Patient also suffers from gastrointestinal problems.
- **Laboratory diagnosis**—histologically, oral epithelial cells in pernicious anemia reveal enlarged, hyperchromatic nuclei with prominent nucleoli and serrated nuclear membranes. The red blood count is decreased often to 3

or less per cubic millimeter. Cells exhibit macrocytosis, poikilocytosis. There is also decreased WBC count and mean corpuscular hemoglobin. In advanced cases of anemia, there are polychromatophilic cells, stippled cells, nucleated cells, Howell-Jolly bodies and Cabot rings. Bone marrow shows great number of immature red cells or megaloblasts with few normoblasts (**Fig. 37-10**), indicating maturation arrest at the more primitive megaloblast state.

- **Gastric pH**—achlorhydria or lack of gastric hydrochloric acid secretion is a constant feature and pH of the gastric content is usually high. Achlorhydria is associated with atrophy of the gastric mucosa, which commonly occurs in presence of chronic inflammation.
- **Schilling test**—Schilling test detects the absence of intrinsic factor. This is discussed in detail in *Chapter 18: Investigations in Dentistry*.

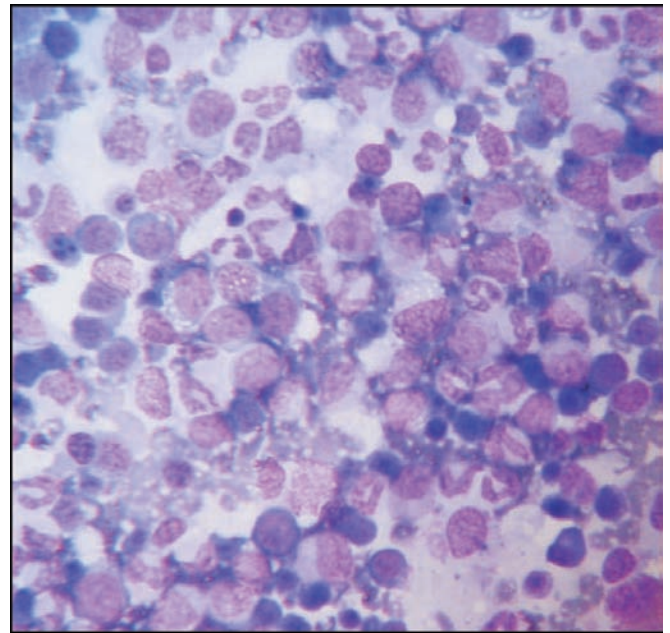


Fig. 37-10: Megaloblasts seen on peripheral smear in patient with pernicious anemia.

Management

- **Vitamin B₁₂**—most of the patients should be given vitamin B₁₂ parenterally but, some of the patients are treated with massive oral dose of vitamin B₁₂. The standard dosage is 100 mg IM every 30 days.

Aplastic Anemia

It is a rare disorder characterized by peripheral blood pancytopenia (anemia, leukopenia and thrombocytopenia) associated with bone marrow suppression. It occurs due to failure of hematopoietic precursor cells in bone marrow to produce adequate number of all types of blood cells. In

most cases, the cause of bone marrow suppression is not known and hence is known as idiopathic aplastic anemia. Aplastic anemia can be associated with Fanconi anemia and dyskeratosis congenita.

Types

- *Primary or idiopathic aplastic anemia*—it is the disease of unknown origin which occurs most frequently in young adults, develops more rapidly and usually terminates fatally.
- *Secondary*—it has known etiology occurs at any age and presents better prognosis.

Etiology

- *Drugs and chemicals*—common drugs which can cause aplastic anemia are benzene derivatives, chloramphenicol, amidopyrine, organic arsenicals, colloidal silver, bismuth, mercury, sulfonamides, penicillin and anticancer drugs. The mechanism due to which drug can cause aplastic anemia is as follows:
 - *Myelosuppressive effect*—the first is characterized by the myelosuppressive effects of cancer chemotherapeutic agents.
 - *Marrow depression*—some drugs can cause marrow depression.
 - *Hypersensitivity reaction*—the third type is called as idiosyncratic or hypersensitivity induced aplastic anemia.
- *Infections*—patient with bacterial disease such as tuberculosis and viral infections like hepatitis and infectious mononucleosis can cause aplastic anemia.
- *Radiation*—long term continuous exposure to small amounts of external radiation may lead to aplastic anemia.
- *Other causes*—in some cases, pregnancy and thymoma can also cause aplastic anemia.

Clinical Features

- *Age*—it can occur at any age, but is common in young adults.
- *Symptoms due to erythrocyte deficiency*—patient may feel weakness after slight physical exertion and exhibits pallor of skin. There is also breathlessness, headache.
- *Symptoms due to platelet deficiency*—there is marked tendency to bruising and bleeding. Patient may get retinal and cerebral hemorrhages.
- *Deficiency of white blood cells*—bacterial and fungal infection are common occurrence due to white blood cell deficiency.
- *Others feature*—patient may get ankle edema, numbness and tingling of extremities, anginal pain, and congestive cardiac failure.

Oral Manifestations

- *Oral mucosa*—the mucosa shows pallor due to decreased number of red blood cells.
- *Symptoms*—in some cases, spontaneous hemorrhage from gingiva is present.
- *Sign*—petechiae, often are present on the soft palate and in severe cases, there may be submucosal ecchymosis. Large ragged ulcers covered by gray or black necrotic membrane may be present, which are the result of generalized lack of resistance to infection and trauma.
- *Gingival hyperplasia*—in some cases, gingival hyperplasia may occur in aplastic anemia (Fig. 37-11).



Fig. 37-11: Gingival hyperplasia associated with aplastic anemia (Courtesy Dr Milind Chandurkar).

Diagnosis

- *Clinical diagnosis*—clinical features suggesting of thrombocytopenia (bleeding), WBC deficiency (infection) and erythrocyte deficiency (pallor) will give clue to the diagnosis.
- *Laboratory diagnosis*—RBC count is remarkably diminished, as low as 1 million cells/mm³. WBC count is as low as 2000/mm³ and platelet count may fall below 20000/mm³. The classical finding is that of pancytopenia along with reduction of absolute reticulocyte count. Bleeding time is prolonged and clotting time is normal. Anemia is normocytic with some degree of macrocytosis. Bone marrow is fatty and few developing cells.

Management

- *Removal of cause*—withdrawal of the etiological agent.
- *Supportive therapy*—it is in the form of antibiotics and transfusion.
- *Hemopoiesis*—stimulation of hemopoiesis by androgenic steroid.
- *Bone marrow transplantation*—it can be useful in some cases.

- *Anti-fibrinolytic agents*—gingival bleeding can be reduced by using systemic anti-fibrinolytic agents such as aminocaproic acid or transexamic acid and local treatment.
- *Immunosuppressive therapy*—it is recommended in patients who are not able to take bone marrow transplantation. Antithymocyte globulin combine with cyclosporine will produce good results.

Fanconi's Anemia

It is a rare congenital disease characterized by pancytopenia, bone marrow hypoplasia and congenital anomalies.

Clinical Features

- *Age*—it is usually seen in children.
- *Skin*—there is brown pigmentation of skin.
- *Hypoplasia*—there is, also, hypoplasia of kidney and spleen.
- *Symptoms*—patient suffers from microcephaly, hypoplastic thumb, and short stature, polydactyly and squint.
- *Other features*—there may be mental and sexual retardation.

Oral Manifestations

- *Gingival bleeding*—there is gingival bleeding which may occur spontaneously.
- *Infection*—frequent infections of oral cavity can occur.

Diagnosis

- *Clinical diagnosis*—pancytopenia with congenital anomalies will diagnose this condition.
- *Laboratory diagnosis*—it is same as that of aplastic anemia.

Management

- *Anti-thrombolytic agents*—gingival bleeding can be reduced by anti-thrombolytic agents such as aminocaproic acid and transexemic acid 20 mg/kg QID 24 hrs before procedure and continued for 3 to 4 days.

Uncommon Causes of Anemia

- *Autoimmune hemolytic anemia*—in this disorder, hemolytic anemia is caused by the presence of antibodies reactive against normal or altered red cell membrane. Antibodies present can be *warm antibody* (IgG antibodies which are active at 37° Celsius) and *cold antibody type* (IgM antibodies below 30° Celsius leading to hemolysis of RBCs).
- *Anemia due to drugs*—number of drugs like naphthalene, nitrofurantoin, amino salicylic acid, phenacetin, dapsone, sulfones and nitrobenzene may cause anemia.

Anemia develops within 1 to 2 weeks after the drug therapy is started and show abnormalities like reticulocytosis, erythrocyte abnormalities, hyperbilirubinemia, and erythroid hyperplasia of the bone marrow.

- *Anemia due to chemicals*—chemicals which can cause anemia are arsenic, potassium chlorate and pyrogallol acid.
- *Anemia due to physical agents*—acute hemolytic anemia has been observed in extensive thermal burns due to direct effect of heat on RBCs, severity of which is related to the area of the body surface affected. Anemia may last for many weeks.
- *Pyruvate kinase deficiency anemia*—when there is pyruvate kinase deficiency during glycolysis in the RBC, there is discrepancy between the ATP generating capacity of RBC and energy needed, leading to loss of pliability which result in distorted and rigid cells which are ultimately destroyed by the spleen. Anemia of varying degrees, jaundice and splenomegaly or pronounced neonatal jaundice are common findings. Anemia is normocytic and normochromic with reticulocytosis.
- *Glucose 6 phosphatase deficiency anemias*—glucose-6-Phosphatase is required to maintain glutathione in the reduced state which protects the RBCs from metha-hemoglobin formation. In normal state there is no anemia but acute hemolysis is characterized by jaundice, pallor and dark urine with or without abdominal and back pain. It remains asymptomatic unless RBCs are subjected to injury by exposure to certain drugs like anti-malarial, vitamin K, aspirin and certain infections like Salmonella, *E. coli*, etc. Peripheral smear shows abnormal forms of RBC and reticulocytosis. There is also increase in WBC count.
- *Hereditary spherocytosis*—in this, there is abnormalities in shape of RBC. In this disorder, red blood cells are excessively permeable to sodium ion. The osmotic fragility of red cells is abnormal in these conditions. It will lead to loss of cell membrane and gradually the cell becomes spherical in shape and is destroyed by spleen, giving rise to hemolytic anemia. Patient will get mild to moderate hepatosplenomegaly, jaundice and anemia. Radiologically, radiating spicule may be present, but they are less widely distributed, although symmetrically situated on two sides of the affected region. The peripheral smear shows presence of spherocytes.
- *Hereditary elliptocytosis*—it is a genetically determined abnormality of the red cell shape associated with variable degree of hemolysis. Most of the patients have no clinical manifestations but few may show signs of chronic hemolytic anemia. The peripheral smear shows larger number of elliptocytes. Mild anemia of normocytic, normochromic type is also present.

- *Paroxysmal nocturnal hemoglobinuria*—it is a very rare disease in which the red cells have acquired susceptibility to antibodies which are normally in the blood. The patient has severe to moderate anemia and mild jaundice. The spleen is usually palpable. There is passage of dark urine usually in morning sample with subsequent samples of urine being clear. There are findings of hemolytic anemia in association with reticulocytosis. There may be leukopenia and thrombocytopenia. Administration of alkali may control hemolysis but on cessation of treatment, massive hemolysis may occur due to accumulation of sensitive cells during the period of therapy.

Dental Considerations of Anemia

- *Blood investigations*—routine blood investigation should be carried out in patient whom you suspecting anemia.
- *Physician referral*—if hemoglobin is significantly less, patient is referred to physician for further diagnosis and treatment.
- *Surgical procedure*—don't perform any surgical and periodontal procedures on patients with marked anemia. Never give general anesthesia unless Hb is at least 10 g/dl.
- *Maintenance of oral hygiene*—oral rinse with chlorhexidine 0.2% in aqueous solution will reduce the amount of plaque and number of microorganisms.
- *Injection*—intramuscular injection and nerve block anesthesia should be avoided because of thrombocytopenia and bleeding tendency.

Polycythemia

It is defined as abnormal increase in the number of red blood cells in the peripheral blood, usually with an increase in hemoglobin level.

Types

- Polycythemia vera
- Relative polycythemia.
- Secondary polycythemia

Polycythemia Vera

It is also called as '*polycythemia rubra vera*', '*Osler's disease*', '*Vaquez's disease*' and '*erythremia*'. There is an uncontrolled proliferation of the erythroid stem cells leading to excess of erythroid cell mass in the body (RBCs). There is accompanying increase in the granulocyte and megakaryocytic elements though to a lesser degree.

Clinical Features

- *Age and sex*—the disease is more common in males and usually occurs in middle age or later.
- *Symptoms*—common symptoms are lassitude, loss of concentration, headache, dizziness and blackout, slurring of speech, pruritis, mental confusion and indigestion. Paresthesia is common, usually involving the cranial nerves.
- *Signs*—the skin appears flushed or diffusely reddened. Spleen is palpable in most of the patients. Superficial veins are dark, enlarged and distended. The tip of the finger usually has a cyanotic appearance.
- *Appearance*—there is marked purplish red discoloration especially of the head and neck (**Fig. 37-12**). Feet and hands, which give the patient an extremely angry appearance.
- *Erythromelalgia*—it is peripheral vascular event occur on hands and feet. There is burning sensation with erythema and warmth in hand and feet. This may lead to thrombotic occlusion of vessels supplying digits which may results in digital gangrene and necrosis.
- *Complications*—thrombotic complications may occur and peptic ulcerations are common.



Fig. 37-12: Purplish red discoloration seen in eyes in patient with polycythemia (Courtesy Dr Millind Chandurkar).

Oral Manifestations

- *Appearance*—there is purplish red discoloration of the ears, oral mucosa, gingiva and tongue.
- *Tongue*—the tongue may appear as if it has been painted with crystal violet.
- *Gingiva*—the gingiva is markedly swollen and bleeds spontaneously, but with no tendency to ulcerations.
- *Oral mucosa*—petechiae of the oral mucosa are common.
- *Hemorrhage*—in such patients, severe hemorrhage may follow surgical procedures.

Diagnosis

- *Clinical diagnosis*—purplish red discoloration of skin with Erythromelalgia accompanied by gingival hemorrhage is typical clue of this disease.
- *Laboratory diagnosis*—the hemoglobin level is greater than 18 gm/dl with an associated elevation of WBC and platelet count. The bone marrow is hypercellular with erythroid hyperplasia, increased number of megakaryocytes and granulocytes.

Management

- *Venesection*—Venesection done at periodic intervals to remove 500-600 ml of blood is the simplest therapeutic measure. It is also called as phlebotomy.
- *Radioactive phosphorus*—radioactive phosphorus may be used when the Diagnosis is certain. It is an excellent treatment modality.
- *Aspirin*—it is given for the thrombotic event.
- *TEM*—oral administration of 30 mg tri-ethylene-melamine (TEM) with remission of 8 to 9 months.
- *Chemotherapy*—chemotherapy with busulfan 2-4 mg/day or melphalan orally to bring down the cell count.

Relative Polycythemia

It is also known as '*stress or spurious (false) polycythemia*' and is due to reduction in the plasma volume.

It is an apparent increase in the number of circulating red blood cells that occurs as a result of loss of blood fluid with hemoconcentration of cells and is seen in cases of excessive loss of body fluids such as chronic vomiting, diarrhea or loss of electrolytes with accompanying loss of water. The red cell mass is normal.

Secondary Polycythemia

This occurs as a result of hypoxia, which stimulates the production of erythropoietin, e.g. at high altitudes, pulmonary diseases or congenital heart diseases.

Secondary polycythemia may also occur in some brain tumors, usually vascular, Cushing's syndrome and renal and lung carcinoma.

White Blood Cell Disorders

For classification of white blood cell disorders (see Table 37-4).

Quantitative Disorders

Agranulocytopenia or Neutropenia

It is also called as '*granulocytopenia*', '*agranulocytic angina*'. It is a serious disease characterized by marked leukopenia

Table 37-4: Classification of white blood cell disorders

Quantitative disorders

Decrease in number

- Granulocytopenia/leukopenia/agranulocytopenia
- Neutropenia
 - Cyclic neutropenia
 - Primary splenic neutropenia
 - Chronic hypoplastic neutropenia
 - Chronic granulocytopenia in childhood
 - Familial neutropenia
 - Transitory neonatal neutropenia

Increase in number

- Granulocytosis
- Neutrophilia
- Basophilic leukocytosis
- Eosinophilic leukocytosis
- Lymphocytosis
- Monocytosis

Qualitative disorders

- Lazy leukocyte syndrome
- Chédiak-Higashi syndrome
- Multiple myeloma
- Infectious mononucleosis
- Lymphoma
- Leukemia

with reduction and absence of neutrophilic leukocytes. Neutropenia is decreases in number of neutrophils and Agranulocytopenia is absent of neutrophils.

Types

- *Primary agranulocytosis*—in this, etiology is unknown.
- *Secondary agranulocytosis*—in it, cause is recognized.
- *Mild neutropenia*—when 1000/mm³ to 2000/mm³ neutrophils are present.
- *Moderate neutropenia*—when 500/mm³ to 1000/mm³ neutrophils are present.
- *Severe neutropenia*—when fewer than 500/mm³ neutrophils are present.
- *Agranulocytosis*—when no neutrophils are seen in peripheral smear.

Etiology

- *Idiosyncrasy*—idiosyncrasy or sensitization to certain drugs like aminophylline, chlorpromazine and phenylbutazone, benzene, bismuth, chloramphenicol, sulfonamides and use of cytotoxic drugs or anti-metabolics.
- *Deficiency*—deficiency of vitamin B₁₂ and folic acid.
- *Infections*—certain infections decrease the number of neutrophils in circulating blood because of increased migration of neutrophils into the tissue, destruction of neutrophils or direct effect of microorganism and its toxins on the bone marrow. Infection with hepatitis A

and varicella zoster virus infection are commonly associated with neutropenia. Overwhelming bacterial infection, particularly septicemia can be accomplished by neutropenia because cells are used at rapid rate to overcome infection.

- *Diseases*—disease causing sequestration of neutrophils includes systemic lupus erythematosus and Felty's syndrome. It can be associated with leukemia, pancytopenia and hypersplenism.
- *Hemodialysis*—hemodialysis patient experience decrease in neutrophils owing destruction by complement activated by the dialysis membrane.
- *Irradiation*—excessive irradiation can cause neutropenia due to direct toxic effect on division of bone marrow cells.

Clinical Features

- *Age and sex*—it can occur at any age but is somewhat more common in adults, particularly in woman.
- *Occupation*—it is also common in professional and in hospital as they have easy access of the offending drugs and often use drug sample injudiciously.
- *Symptoms*—the onset may be sudden or gradual. The condition begins with sore throat, high fever and often rigors, which may be followed by prostration.
- *Skin*—the skin appears pale and anemic and in some cases, jaundiced.
- *Signs*—there is rapidly advancing necrotic ulceration of throat and mouth with little evidence of pus formation. In case of Agranulocytopenia patient dies within 3-5 days due to toxemia and septicemia.

Oral Manifestation

- *Sites*—the most common sites are gingiva (**Fig. 37-13**), palate, tonsil and pharynx.



Fig. 37-13: Patient having severe gingivitis with neutropenia.

- *Symptoms*—There may be associated pain, excessive salivation and spontaneous oral hemorrhage.
- *Appearance*—the oral lesions appear as necrotizing ulcerations of oral mucosa, tonsils or /and pharynx.
- *Surface of the lesion*—the lesions appear as ragged and necrotic and are covered with a gray black membrane. The necrotic tissue is often with foul smelling.
- *Margins of lesion*—there is lack of inflammation at the margin of lesions.
- *Supporting structures*—the disease spreads quickly in gingival tissues causing destruction of supporting structures and inevitable loss of deciduous and permanent teeth.

Radiological Findings

- *Destruction of alveolar bone*—supporting alveolar bone is rapidly destroyed, so that teeth are denuded of bone and are supported only by soft tissues.
- *Osteomyelitis*—very rarely, infection spreads to deep into the marrow to cause osteomyelitis.

Diagnosis

- *Clinical diagnosis*—gingival ulceration, infection and anemic patient may give clue to the diagnosis
- *Laboratory diagnosis*—majority of patients show a leukocyte count below 2000 cells per cu mm and granulocyte count below 100 cells per cu mm. Hemoglobin and platelet counts are normal.

Differential Diagnosis

- *Leukemia*—anemia, purpura and WBC leukocyte above 1000,000/mm³ with prolonged bleeding and clotting time.
- *Cyclic neutropenia*—marked cyclic appearance every 20 to 25 days.
- *Wegener's granulomatous*—kidney and lung involvement.
- *ANUG*—necrosis starts on papilla tips, not necessary associated with leukopenia.
- *Necrotizing sialometaplasia*—large ulcerations with induration on hard and soft palate, flat border and painless. General condition is not affected.
- *Erythema multiforme*—target lesions and no specific blood picture.

Management

- *Removal of causative agent*—the most important measure is removal of the offending agent if it can be identified.
- *Transfusion*—if hemoglobin is less than 10 gm/dl, transfusion of red cell concentrate is given. In some cases, white cell transfusion can be given.

- **Antibiotics**—septicemia can be controlled by parenteral antibiotic therapy along with corticosteroid therapy. A combination of carbenicillin, methicillin and gentamicin is commonly used because of broad coverage against most organisms.
- **Granulocyte macrophage colony stimulating factors (GM-CSF)**—this is given if agranulocytosis is related to cancer treatment.
- **Oral and dental management**—antibacterial mouth rinse may be helpful for ulcers. The soft splint which covers entire maxillary area and carries a medication is used in palatal ulcers. The pain of oral ulcer is reduced by use of topical anesthetic mouth rinses. A solution containing 5% dyclonine and 5% diphenylhydramine hydrochloride (benydryl) mixed with magnesium hydroxide (milk of magnesia) or kaolin with pectin is useful for this purpose. Use of splint to cover the palatal lesion and carries medication in a well that continually bathes the oral ulcers.

Cyclic Neutropenia

It is also called as '*periodic neutropenia*' or '*cyclic hematopoiesis*'. It is a rare disorder characterized by periodic or cyclic diminution in circulating neutrophils due to failure of stem cells of bone marrow. It is also called as periodic neutropenia. One-third cases are inherited as autosomal dominant trait and 2/3rd appear spontaneously during the first few year of life. The patient is healthy between neutropenic periods; but at regular intervals, the absolute neutrophils count falls below $500/\text{mm}^3$. In some patients, it comes to zero.

Clinical Features

- **Age and sex**—the disease is frequently present during infancy and childhood and both sexes appear to be equally affected.
- **Occurrence**—the frequency of neutropenic episodes vary from once in 2 to 4 weeks which last for 3 to 5 days with 21 days gap being the commonest.
- **Symptoms**—the patients manifest fever, sore throat, stomatitis and regional lymphadenopathy as well as headache, arthritis, cutaneous infection and conjunctivitis. Less frequently patient experience lung, urinary tract infection as well as rectal and vaginal ulcer.
- **Amyloidosis**—in some patients, amyloidosis can occur due to repeated, increased antigenic stimulation during neutropenic episodes.

Oral Manifestations

- **Site**—lesions are found on the lip, tongue, palate, gums and buccal mucosa.

- **Gingivitis**—severe gingivitis is present on gingiva.
- **Ulceration**—painful ragged ulcers that have a core like center are present. It heals after about two weeks with scarring. Isolated painful ulcer may occur which correspond to the period of neutropenia.

Radiological Features

- **Bone loss**—it exhibits mild or severe loss of superficial alveolar bone advancing to periodontitis.
- **Prepubertal periodontitis**—in children this loss of bone around multiple teeth sometimes called as '*prepubertal periodontitis*'.

Diagnosis

- **Clinical diagnosis**—recurrent gingival ulceration, recurrent infection with prepubertal periodontitis may yield in diagnosis.
- **Laboratory diagnosis**—neutrophils count should be less than $500/\text{mm}^3$ during each of three successive cycles. There is increased in monocytes and lymphocytes count. At the peak of the disease, the neutrophils may completely disappear for one or two days.

Management

- **Antibiotic therapy**—this can be given to control the infection in the patient.
- **Corticosteroid therapy and hormonal therapy**—in some cases, use of corticosteroids, adrenocorticotropin (ACTH) or testosterone modulates sharp reduction in marrow function.
- **Oral hygiene maintenance**—oral hygiene should be maintained and patient should be recalled for oral hygiene every 2-3 months.
- **Cytokine granulocyte colony-stimulating factor (G-CSF)**—this can be given several times weekly to correct lack of production of neutrophils. This will decrease time of neutropenia from 5 days to 1 day thereby decreasing clinical course of disease.
- **Other therapy**—other therapy like splenectomy and nutritional supplement can be carried out.

Variant of Neutropenia

- **Chronic idiopathic neutropenia**—it is an uncommon disease in which there is decreased production of neutrophils. The bone marrow shows a number of immature cells but there is a decrease in number of mature neutrophils. Most of the patients are asymptomatic because of compensatory monocytosis. In the oral cavity recurrent ulcerations of the gingival, lips, buccal mucosa and recurrent dental abscess have been reported.

- *Primary splenic neutropenia*—it is characterized by neutropenia of a mild degree, splenomegaly and myeloid hyperplasia of the bone marrow. The common clinical features are polyarthritis, recurrent oral ulceration, weight loss and fatigue.
- *Chronic granulocytopenia in children*—it is a benign disorder characterized by repeated pyogenic infection. It is due to increased destruction of neutrophils. Complete recovery takes place eventually.
- *Familial neutropenia*—it is a rare benign disorder transmitted as a non-sex linked dominant trait. Most of the patients are asymptomatic but a few may show periodontal disease and a tendency to develop frequent furuncles. The total leukocyte count is normal but there is associated neutropenia.
- *Transitory neonatal neutropenia*—it is a disorder caused by fetal-maternal incompatibility involving only the neutrophils. Skin infection is the most common clinical finding. The blood picture shows normal leukocyte count, severe neutropenia, monocytosis and mild eosinophilia. The duration of neutropenia varies from 2 to 17 days.

Increased in Number of WBC

- *Neutrophilia*—it is characterized by an increase in blood neutrophil concentration above 7.25×10^6 . It is seen in acute and chronic infection.
- *Granulocytosis*—in this, there is an increase in the granulocytes and is seen in patients with chronic infection, Hodgkin's disease, polycythemia vera and skin inflammation or any activity that increase epinephrine release such as stress or exercise.
- *Eosinophilia*—it is an increase in number of eosinophilic leukocytes and is seen in allergic reactions, skin disease, parasitic infection, leukemia, polycythemia, malignancy, and following irradiation.
- *Basophilic leukocytosis*—an increase is seen in myxedema, ulcerative colitis, chronic sinusitis, leukemia, polycythemia, Hodgkin's disease and following splenectomy.
- *Monocytosis*—it is seen in bacterial infection, recovery from acute infection and agranulocytosis, protozoal infection, lymphoma, leukemia, carcinoma of stomach breast cancer and high dose of steroid therapy.
- *Lymphocytosis*—it is seen in certain acute infections like brucellosis, tuberculosis, syphilis, leukemia and lymphosarcoma.

Qualitative Disorders of WBC

Lazy Leukocyte Syndrome

It is also called as '*Schwachman syndrome*'. It is a result of loss of chemotactic function of the neutrophil. The

neutrophils present in the blood cannot migrate to the site of tissue injury although phagocytic and bactericidal activities are normal.

Clinical Features

- *Age*—it becomes apparent at the age of 1-2 years when complication occurs due to infections.
- *Symptoms*—the most common clinical manifestations are stomatitis, otitis media and bronchitis.
- *Recurrent infection*—chances of recurrent infection in the patients are high.

Oral Manifestations

- *Stomatitis*—it is common oral finding of this disorder.
- *Periodontitis*—in some cases, periodontal disease may be present.

Diagnosis

- *Clinical diagnosis*—recurrent infection, periodontitis and stomatitis may give clue to diagnosis.
- *Laboratory diagnosis*—the total leukocyte count is slightly low but the absolute neutrophil count is as low as 100 to 200 cells/mm³. The bone marrow contains normal number of mature neutrophils.

Management

- *Antibiotics*—antibiotics are given to control the infection.

Chédiak-Higashi Syndrome

It is a congenital autosomal recessive defect of granulocytes and melanocytes. Abnormal granules are seen in all blood granulocytes resulting in decreased chemotactic and bactericidal activity.

Clinical Features

- *Albinism*—the characteristic clinical feature of this disease consists of oculocutaneous albinism.
- *Recurrent infection*—patient with Chediak Higashi syndrome will have recurrent infections of the respiratory tract and sinuses.
- *Gastrointestinal disturbance*—patient is having hepatosplenomegaly.
- *Lymph node enlargement*—there is cervical lymph node enlargement present in this patient.
- *Malignant lymphoma*—the disease may be associated with malignant lymphoma.
- *Other diseases*—there is photophobia, nystagmus, thrombocytopenia, and neurological problems.

Oral Manifestations

- *Ulceration*—ulceration of the oral mucosa is very common.
- *Periodontitis*—There may be loss of teeth due to periodontal disease.
- *Gingivitis and glossitis*—inflammation of gingiva and tongue are common occurrence.

Diagnosis

- *Clinical diagnosis*—albinism, recurrent infection, hepatosplenomegaly, oral ulceration and periodontitis will give clue to the diagnosis.
- *Laboratory diagnosis*—hematological studies show presence of giant abnormal granules in the granulocytes in the peripheral blood as well as in their precursors in the bone marrow.

Management

- *Antibiotics*—Immediate and proper treatment with antibiotics of the infection as soon as they occur is most important.
- *Other drugs*—vincristine, prednisolone and ascorbic acid have been tried as the treatment of this disorder.

Multiple Myeloma

It is described in Chapter 16: Malignant Tumor of Jaw.

Lymphoma

It is described in Chapter 16: Malignant Tumor of Jaw.

Leukemia

It is described in Chapter 16: Malignant Tumor of Jaw.

Chronic Idiopathic Neutropenia

It consists of group of diseases which includes familial neutropenia, chronic benign neutropenia, chronic neutropenia and hyperplastic neutropenia. Etiology is unknown, but they are characterized by a decrease in production of neutrophils in the bone marrow.

Clinical Features

- *Age and sex distribution*—it is seen in young adults and seen in predominantly in females.
- *Symptoms*—it is usually asymptomatic and free of infection which is due to compensatory monocytosis, which accounts of normal number of phagocytes at the site of tissue injury. In some cases recurring upper respiratory tract infections, otitis media, bronchitis and furunculosis can also occur.

Oral Manifestations

- *Periodontitis*—there is severe, rapidly advancing periodontal diseases.
- *Gingival finding*—gingivae appear intensely red with granulomatous margins. Severe gingival recession, advanced bone loss, mobility, denuded roots and loss of teeth.
- *Recurrent oral ulceration*—the second most common finding is recurring oral ulcers and in some cases, maxillary sinusitis can also occur.

Diagnosis

- *Clinical diagnosis*—it is not so specific.
- *Laboratory diagnosis*—the bone marrow of patients shows a normal number of immature cells but decrease in number of mature neutrophils. This is called as maturation arrest. Absolute neutrophils count may be below 500/mm³.

Management

- *Corticosteroids*—it is not required if the patient is asymptomatic. Patient who develops severe recurrent infections may benefit from alternate day corticosteroids therapy.

Platelet Disorders

Classification

Purpura

- Idiopathic thrombocytopenic purpura
- Secondary thrombocytopenic purpura
- Congenital purpura
- Thrombotic thrombocytopenic purpura
- Thrombocytopathic purpura
 - von-Willebrand's disease
 - Bernard-Soulier syndrome
 - Aldrich syndrome

Thrombocytosis

Thrombocytopenic Purpura

Purpura is defined as a purplish discoloration of the skin and mucous membrane due to subcutaneous and submucous extravasation of blood. Purpura may be due to deficient or defective platelets or due to an unexplained increase in capillary fragility.

It is also called as 'Werlhof's disease', 'purpura hemorrhagica' and 'primary thrombocytopenic purpura'. It is a disease in which there is an abnormal reduction in the number of circulating blood platelets with normal or raised number of megakaryocytes in the bone marrow. It is thought to be

an autoimmune disorder in which a person becomes immunized and develops antibodies against his own platelets. It can be a result of decrease in production of platelets or increased destruction of platelets or, both. Clinical features occur if platelet counts drop below $100,000/\text{mm}^3$.

Etiology

- **Idiopathic**—in some cases etiology is unknown causing the decrease in platelet production.
- **Reduced production**—it occurs due to infiltration of bone marrow by malignant cells. It can occur due cancer chemotherapeutic drugs.
- **Platelet destruction**—there may be increased in platelet destruction due to immunological causes. This can be caused by many drugs like heparin.
- **Increase consumption**—increase in consumption of platelet may occur in abnormal blood clot formation. This is seen in thrombotic thrombocytopenic purpura.
- **Spleen sequestration**—condition in which splenomegaly is present, large number of platelet are taken out for circulation.
- **Systemic disease**—some systemic disease like lupus erythematosus and HIV infection may cause purpura. Other diseases like leukemia, sarcoma, lymphoma, Hodgkin's disease myelofibrosis can also cause purpura.
- **Drugs**—barbiturates, analgesics (phenylbutazone and salicylates), antihistamine (diphenhydramine hydrochloride), myelosuppressive agents used in neoplastic disease.
- **Metabolic disease**—uremia, megaloblastic anemia can also cause purpura.
- **Others**—bone marrow replacement, radiation therapy can also cause purpura.

Clinical Features

- **Petechiae**—thrombocytopenic purpura is characterized by spontaneous appearance of purpuric hemorrhagic lesions of skin. The reason for this capillary is not able to seal off with thrombi. The patient also exhibits a bruising tendency.
- **Ecchymosis**—if large quantity of blood is extravasated Ecchymosis may result (**Fig. 37-14**).
- **Hematoma**—this can occur in rare case. It also occurs due to large amount of blood extravasated, which vary in size from tiny red pinpoint petechiae to large purplish ecchymosis and sometimes, even massive hematoma.
- **Other features**—epistaxis, hematuria and melena are common findings.
- **Intracranial hemorrhage**—intracranial hemorrhage is rare, but can be seen in children and the symptoms are headache, dizziness and confusion.



Fig. 37-14: Ecchymosis seen on hand in patient with thrombocytopenia purpura.

- **Congenital purpura**—it may result from placental transfer of platelet antibodies. Such antibodies may arise from active immunization of the mother by fetal antigens or from passive transfer of auto-antibodies present in the maternal circulation. In infants, it is first seen at about one to two weeks of age. This disorder is frequently mild or even subclinical. Some cases show generalized petechiae and ecchymosis. In some infants, hemorrhage can be severe and intracranial hemorrhage may result in death. As the responsible antibodies disappear from the circulation platelet count returns to normal within 1 to 4 weeks.
- **Thrombotic thrombocytopenic purpura**—it is immunologically mediated. It is characterized by occlusion of small arterioles and capillaries of many organs by thrombi formed of fibrin and platelets. It generally occurs in young adults and more commonly in females than in males. There is thrombocytopenia, hemolytic anemia, fever, transitory neurologic dysfunction and renal failure.

Oral Manifestations

- **Site**—submucous petechiae and ecchymosis commonly occur especially at the junction of the hard and soft palate (**Fig. 37-15**). In some cases, it can be seen on tongue.
- **Bleeding after tooth extraction**—the first manifestation of the disease can be seen in oral cavity in the form of excessive bleeding after tooth extraction.
- **Petechiae**—it appears as numerous tiny, grouped clusters of reddish spots, only a millimeter or less in diameter. Petechiae do not blanch on pressure which is the distinguishing feature between purpura and telangiectasia.
- **Gingival bleeding**—in severe cases, extensive spontaneous gingival bleeding may be seen and this may form foci of secondary infection.
- **Tongue**—in some cases, petechiae and ecchymosis can be seen on the tongue (**Figs 37-16A and B**).



Fig. 37-15: Thrombocytopenic purpura showing petechiae at the junction of hard and soft palate.



Figs 37-16A and B: Petechial hemorrhage seen on tongue in patient with thrombocytopenic purpura.

Diagnosis

- **Clinical diagnosis**—petechiae at the junction of hard and soft palate is clue to the diagnosis (**Fig. 37-17**).

- **Laboratory diagnosis**—the platelet count is usually below 60,000 cells per cu mm. The bleeding time is prolonged but, the clotting time is normal. The bone marrow reveals megakaryocytic hyperplasia. When severe bleeding occurs, there may be associated iron deficiency anemia.



Fig. 37-17: Petechiae seen at the junction in hard palate area as pinpoint purplish lesion.

Management

- **Young children**—in young children, treatment is seldom necessary unless there is intracranial evidence of bleeding.
- **Corticosteroids**—in adults, prednisone 60 mg/dl should be given until the platelet count rises to normal level when, the dose should be cautiously tapered until the drug is withdrawn.
- **Splenectomy**—if there is no response to prednisolone in 3 to 4 days, splenectomy is indicated.
- **Transfusion**—platelet transfusion can be given.
- **Removal of cause**—in case of secondary thrombocytopenia purpura, this will reduce the symptoms and sign of lesion.
- **Local hemostasis**—the spontaneous gingival hemorrhage can often be controlled by the local use of hemostatic of the non-causing type, such as fibrin foam, Gelfoam or absorbable cellulose with thrombin. Sometimes, 1.5% hydrogen peroxide mouth wash will stop gingival bleeding.

Von Willebrand's Disease

It is also called as '*pseudohemophilia*', '*vascular hemophilia*' and '*vascular purpura*'. It is rare disorder which is inherited as an autosomal dominant trait in which there is defect in all three components of the hemostatic mechanism; the capillaries, platelets and coagulation mechanism. It is most common hereditary clotting disorder. In it, there is

quantitative and qualitative deficiency in larger molecule portion of factor VIII molecule. Von Willebrand's factor activity is responsible for platelet adhesion to subendothelium, regulation of the plasma level of factor VIII coagulant activity and thereby normal hemostasis.

Clinical Features

- *Age and sex distribution*—it is seen in childhood and adults and females are affected more commonly than males.
- *Bleeding tendencies*—excessive bleeding either spontaneously or following even minor trauma is the chief feature of the disease. Bleeding into the gastrointestinal tract, epistaxis and severe menorrhagia are also common. The most common sites of bleeding are nose, skin and gingiva. Bleeding tendencies usually appear early in childhood and decrease in middle and old age.

Oral Manifestations

- *Gingival bleeding and post-extraction bleeding*—gingival bleeding and post-extraction bleeding are the most common oral manifestations. The disease may be discovered after dental extraction.

Diagnosis

- *Clinical diagnosis*—bleeding tendencies either spontaneous or after slight trauma may give clue to the diagnosis.
- *Laboratory diagnosis*—there is prolonged bleeding time, normal platelet count, failure of aggregation of platelets impaired adherence of platelets and depressed levels of factor VIII are suggestive of this disorder. There is low Anti-hemophilic globulin (AHG) levels and abnormal platelet retention to glass beads. The platelet count and prothrombin consumption are normal. The clotting time usually is normal but capillary fragility is increased.

Management

- *Transfusion*—Bleeding episode is best controlled by transfusion of plasma and or cryoprecipitate and by local control of homeostasis.
- *Avoid giving aspirin*—patients with von Willebrand's disease should not be given aspirin because of prolongation of the bleeding time that may occur.
- *Desmopressin*—it is given as nasal spray to control nasal bleeding.

Aldrich Syndrome

It is also called as 'Wiskott-Aldrich syndrome'. It is X-linked recessive condition. It is characterized by thrombocytopenia.

Clinical Features

- *Symptoms*—patients commonly manifest boils, otitis media, bloody diarrhea and respiratory infection.
- *Increased susceptibility to infection*—there is eczema and increased susceptibility to infection.
- *Malignant lymphoma*—there is common occurrence of malignant lymphoma, which is an important feature of this disease.
- *Bleeding*—bleeding occurs from nose, skin and gastrointestinal tract.

Oral Manifestations

- *Palatal petechiae*—palatal petechiae are frequently present.
- *Spontaneous bleeding*—there is spontaneous bleeding from the gingiva.

Diagnosis

- *Clinical diagnosis*—increase susceptibility to infection, spontaneous bleeding, and malignant lymphoma may give clue to diagnosis.
- *Laboratory diagnosis*—prolonged bleeding time, anisocytosis, alteration in the size and shape of platelets with most platelets smaller than normal. There is decreased production and defective maturation of platelets since normal megakaryocytes may be seen in the marrow.

Management

- *Antibiotics*—patients are given antibiotics to control the infection.
- *Hemostasis*—local hemostasis to control gingival bleeding should be done.

Familial Thrombasthenia

It is also called as 'Glanzmann's disease'. It is hereditary, chronic hemorrhagic disease transmitted as an autosomal recessive trait. There is defective platelet aggregation due to defective membrane protein.

Clinical Features

- *Sex distribution*—both sexes may be affected and onset of menarche may be a critical event.
- *Severe bleeding*—there is excessive bleeding, either spontaneous or following minor trauma.
- *Sign and symptoms*—purpuric hemorrhages of skin are common, as are epistaxis and gastrointestinal bleeding. Hemarthrosis is also noted in some cases.

Oral Manifestations

- *Spontaneous gingival bleeding*—spontaneous bleeding from the oral cavity particularly gingival bleeding.

Diagnosis

- *Clinical diagnosis*—severe excessive bleeding, epistaxis and hemarthrosis can be present.
- *Laboratory diagnosis*—bleeding time is prolonged while clot retraction is impaired. Platelet count is normal as the clotting time. The aggregation of platelet by epinephrine, ADP and thrombin is defective.

Management

- *Platelet infusion*—no specific treatment but patient in oral surgery can be given microfibrillar collagen preparation with fibrinolytic inhibitors.

Onyiai

It means blood blister. It is variant of idiopathic thrombocytopenic purpura because of its similarities to hematological manifestations. It is thought to be linked to the traditional diet and lifestyles prevalent in primitive societies of Southern and East African countries.

Clinical Features

- *Age and sex distribution*—individuals in the second decade of life seen most commonly affected although, the disease can develop any time after the first 6 months of life. There is female predilection.
- *Onset*—the sudden appearance of hemorrhagic bullae on the skin and oral mucosa followed by epistaxis or frank bleeding from mouth.
- *Signs and symptoms*—there is also subconjunctival hemorrhages, hematemesis, melena, hematuria, vaginal bleeding and CNS hemorrhage. Hemorrhagic episodes may be multiple and recurrent attacks result in anemia.

Diagnosis

- *Clinical diagnosis*—sudden hemorrhagic bulla appearance will give clue to diagnosis.

Management

- *Drug management*—corticosteroid therapy, blood transfusion and platelet transfusion

Thrombocytosis or Thrombocythemia

It is characterized by an increase in the number of platelets in the blood in excess of 1000×10^6 /dl. It is of two types, i.e. primary and secondary.

It is caused by polycythemia and myeloid leukemia, anemia, tuberculosis, sarcoidosis, hyperadrenalism, rheumatoid arthritis, and bronchial carcinoma.

Clinical Features

- *Age*—it is a rare disorder of the elderly associated with a tendency to bleed and to have thrombotic episodes.
- *Epistaxis*—epistaxis, bleeding into the gastrointestinal tract as well as bleeding into the genitourinary tract and central nervous system is common.
- *Skin hemorrhage*—hemorrhages in skin are also common.

Oral Manifestations

- *Gingival bleeding*—spontaneous gingival bleeding and excessive and prolonged bleeding after extraction of tooth is also common.

Diagnosis

- *Clinical diagnosis*—epistaxis and gingival bleeding will give clue to diagnosis.
- *Laboratory diagnosis*—the platelet count is increased and there is abnormal aggregation in response to several aggregating agents. The clotting time and prothrombin time are normal but bleeding time is frequently prolonged.

Management

- *Drug management*—certain cytotoxic drugs, heparin, corticosteroid and aspirin in small dose may help to counteract the thrombotic tendency.
- *Radioactive phosphorus*—it responds to radioactive phosphorus.

Hemorrhagic Disorders

Hemophilia

Hemophilia stands for hemo (blood), philia (loving). It is a hereditary disorder of blood coagulation characterized by excessive hemorrhage due to a prolonged coagulation time. Deficiency of factor VIII or anti-hemophilic factor is the cause of hemophilia. It is transmitted as X-linked recessive character carried on X-chromosome. The males are clinically affected and the females are carriers of the trait. Hemophilia A occurs 10 times more commonly than hemophilia B. Traumatic injury of the oral cavity may lead to the diagnosis of hemophilia.

Hemophilia B is also called as 'Christmas disease'. It occurs due to hereditary deficiency of factor IX or functionally defective factor IX. It is transmitted as X-linked recessive character through chromosome. It is very rare, compared to hemophilia A.

Types (according to AHG factors)

- *Mild*—less than 4% of AHG

- *Moderate*—1% to 3% of AHG
- *Moderate to severe*—0.0% to 0.9% of AHG
- *Severe*—0% of AHG

Clinical Features

- *Age and sex distribution*—these manifestations usually begin after 6 months of age, when the child begins to move about and tends to fall and sustain injuries, i.e. when spontaneous hemorrhage is noted by the parents.
- *Joint hemorrhage or hemarthrosis*—the most common manifestation is hemorrhage into joints which is spontaneous and associated with warmth and muscle spasm. Repeated episodes cause damage to the joint with wasting of the related muscle, leading to deformity and crippling.
- *Subcutaneous hemorrhage*—hemorrhage into subcutaneous tissues, internal organs and musculature also are frequent and potentially disabling complications. Superficial trauma gives rise to uncontrolled bleeding.
- *Intracranial bleeding*—intracranial bleeding is relatively rare unless associated with trauma.
- *Pseudotumor of hemophilia*—in some cases tissue hemorrhage can lead to formation of tumor-like mass.

Oral Manifestations

- *Site*—the anatomic sites involved in persistent oral bleeding are the frenum of lip and the tongue.
- *Symptoms*—there is prolonged bleeding after tooth extraction.
- *Hematoma of floor of mouth*—hematoma of the floor of mouth may occur and blood may spread via the fascial spaces and produce a hematoma of the larynx, with consequent respiratory embarrassment.
- *Tooth eruption*—physiological processes of tooth eruption and exfoliation may be associated with severe and prolonged hemorrhage.
- *Gingival hemorrhage*—gingival hemorrhage may as a result of gingival injury (Fig. 37-18).

Diagnosis

- *Clinical diagnosis*—extensive bleeding after extraction of tooth and routine periodontal procedure should suspect hemophilia.
- *Laboratory diagnosis*—clotting time is prolonged, but however the bleeding time, platelet count and prothrombin time are all normal. The prothrombin consumption time and the partial thromboplastin time may be prolonged in severe cases.
- *Difference between hemophilia A and hemophilia B*—in the laboratory, hemophilia A may be differentiated from hemophilia B by modification of the prothrombin consumption time or the partial thromboplastin time.



Fig. 37-18: Gingival hemorrhage seen in hemophilia patient.

Management

- *Fresh frozen plasma*—replacement therapy of fresh frozen plasma should be given. Hypovolemia, allergic reaction, transfusion hepatitis, hemolytic anemia and development of factor VIII antibodies are complications with fresh frozen plasma.
- *Factor VIII concentrates*—the dose and duration of administration of factor VIII concentrates are planned, taking into consideration of the site and the type of bleeding.
- *Prevention of complication*—to reduce complications caused by cryoprecipitate or plasma various drugs are used instead of factor VIII products, before dental extraction, e.g. a synthetic analogue of anti-diuretic 1-deamino-8-d-arginine vasopressin (DDAVP) in combination with tranexamic acid and Epsilon aminocaproic acid (EACA) can be given.
- *Genetic counseling*—it should be provided to patient to know importance of inheritance.

Dental Management of Hemophilia A and Hemophilia B

- *Anesthesia*—local anesthesia is contraindicated in severe hemophiliac patients with prior replacement therapy. If to be given, it should be intrapulpal anesthesia, intra-ligamentary (periodontal) and papillary injection. Sedation with diazepam or NO₂/O₂ analgesia can be given.
- *Endodontic therapy*—common endodontic procedure is quite acceptable provided care is taken not to exceed beyond the apex of tooth. Hemorrhage into the canal can usually be controlled with 1:1000 aqueous epinephrines on an endodontic paper point.
- *Restorative*—it can be carried out in hemophiliac patient, but the rubber dam must be used to prevent trauma to

gingiva and other soft tissues. When use of rubber dam is not practical, an epinephrine impregnated hemostatic cord is placed in the gingival sulcus before preparation of the crown or inlay margins.

- *Prosthodontics therapy*—complete dentures are well tolerated in hemophilic patients. Partial dentures are also well tolerated, as long as the patient maintains meticulous oral hygiene because clasp can trap food debris, resulting in gingivitis and subsequent hemorrhage.
- *Periodontic therapy*—conservative periodontal treatment is generally more desirable than gingival and osseous surgery because of hospitalization and extensive amount of replacement therapy necessary with surgery.
- *Oral surgical procedure*—Local homeostatic agents and techniques include pressure surgical packs, oxidized cellulose saturated with bovine thrombin solution, vasoconstrictors, suture, topical thrombin and use of absorbable homeostatic. After packing, the tooth socket must be protected by mechanical splint to prevent disturbance to the clot. Postoperative use of anti-fibrinolytic agent and adherence to soft diet are further advocated to support clot maintenance.
- *Drugs precaution*—aspirin and other NSAIDs are avoided in patients with bleeding disorders owing to inhibition of their platelet function. Intramuscular injection should also be avoided because of risk of hematoma formation.
- *Electrosurgery*—electrosurgery is not recommended because it causes tissue necrosis and possible subsequent hemorrhage.

Factor V Deficiency or Parahemophilia

It is a rare hemorrhagic disorder, clinically similar to hemophilia, caused by a deficiency of factor V or proaccelerin.

Clinical Features

- *Age and sex distribution*—it is inherited as an autosomal dominant trait affecting both sexes.
- *Symptoms*—spontaneous epistaxis, bleeding into the gastrointestinal tract and menorrhagia are common.
- *Cutaneous lesion*—cutaneous ecchymosis and hamartomas are frequently seen, although petechiae are rare.
- *Others*—intraocular hemorrhage and hemorrhage into the central nervous system have been also reported.

Oral Manifestations

- *Gingival bleeding*—spontaneous gingival bleeding occurs in some cases and prolonged bleeding after extraction of tooth is observed.

Diagnosis

- *Clinical diagnosis*—spontaneous epistaxis, intraocular hemorrhage with gingival bleeding.
- *Laboratory diagnosis*—both clotting and prothrombin time are prolonged but, bleeding time is normal. The basic defect is reduction in plasma proaccelerin.

Management

- *Transfusion*—transfusion and freshly frozen plasma are given when there is excessive hemorrhage.

Hypofibrinogenemia

It is uncommon disease in which patient has little or no fibrinogen present, in either his plasma or tissue.

Types

- *Congenital*—it is rare and autosomal recessive trait.
- *Acquire*—it occurs secondary to increase in fibrinogen consumption during intravascular clotting, defective fibrinogen formation and destruction or digestion of fibrinogen by fibrinolytic or proteolytic enzymes circulating in the bloodstream.

Clinical Features

- *Sex*—it occurs in both sexes with slight predilection for males.
- *Symptoms*—patients exhibit severe bleeding episodes throughout their lives and the disease is clinically indistinguishable from hemophilia. There is also epistaxis, bleeding into gastrointestinal tract and central nervous system has been also reported.
- *Cutaneous lesion*—cutaneous ecchymosis and hamartomas are frequently seen, although petechiae are rare.

Oral Manifestations

- *Gingival bleeding*—spontaneous gingival bleeding and prolonged bleeding, after extraction of tooth is observed.

Diagnosis

- *Clinical diagnosis*—it is not possible to differentiate it from hemophilia clinically.
- *Laboratory diagnosis*—bleeding time may be normal or slightly prolonged but prothrombin time and clotting time are infinite.

Management

- *Transfusion*—there is no specific treatment for the disease except for transfusion, particularly of concentrated fibrinogen during bleeding episodes.

Macroglobulinemia

It is also called as 'macroglobulinemia of Waldenstrom'. It is a plasma cell dyscrasia showing hemorrhagic tendency similar to other hemorrhagic diseases. The formation of complex between the abnormal immunoglobulin M, macroglobulin and clotting factors, thereby inactivating the clotting factors is responsible for hemorrhage.

Etiology

- *Multiple myeloma*—it is a variant of multiple myeloma.
- *Bing-Neel syndrome*—Bing-Neel syndrome (hyperglobulinemia with central nervous system involvement on toxic-infectious basis).
- *Plasmacytoma*—it is also a variant of plasmacytoma.
- *Immunological*—it is an altered immunologic basis.

Clinical Features

- *Age and sex distribution*—it is most common in persons over age of 50 years, seldom under 40 years and males and female affected equally.
- *Signs and symptoms*—the chief clinical signs are weakness, weight loss, pallor, lymphadenopathy, hepatomegaly, epistaxis, subarachnoid, and ocular hemorrhage.

Oral Manifestations

- *Spontaneous gingival hemorrhage*—it consists of spontaneous gingival hemorrhage, often with continued oozing of blood.
- *Oral ulcer*—bleeding oral ulcers on tongue, palate, buccal mucosa or gingiva and focal areas of hyperemia which appear edematous are painful.

Diagnosis

- *Clinical diagnosis*—oral ulcer, gingival bleeding, lymphadenopathy, weakness, and hepatomegaly will give clue to diagnosis.
- *Laboratory diagnosis*—it generally manifests severe anemia with hemoglobin level near 4 to 6 gm/dl. An extremely elevated sedimentation rate with demonstrable euglobulins and frequent gelling of the serum upon cooling to room temperature or lower are cooling. The viscosity of blood serum is high. The WBC count, clotting time, bleeding time and prothrombin time are normal but lymphocytosis, neutropenia and thrombocytopenia are occasionally seen.

Management

- *Chlorambucil*—chlorambucil, in high doses, provides prolonged remission in these patients.

- *Plasmapheresis*—repeated plasmapheresis has often been used for temporary treatment.

Hereditary Hemorrhagic Telangiectasia

It was first described by Osler in 1901 and it frequently bears his name (*Osler-Rendu-Weber syndrome*). It is transmitted as an autosomal dominant trait and characterized by bleeding from mucous membrane and telangiectatic lesions on skin and mucosa. Telangiectasia represents permanently enlarged capillaries that are localized superficially, just under the skin and mucosa.

Clinical Features

- *Age*—the lesion may be present in childhood but more often appear in puberty and become progressively worse with increasing age.
- *Site*—they are found most commonly on the skin of the face, finger, toes, and on the oral mucous membrane.
- *Appearance*—the disease is characterized by multiple localized angiomas or telangiectases on the skin.
- *Signs*—the lesion bleeds easily, even after slightest trauma. Bleeding is not caused by clotting factor deficiency but as a result of rupture of the weak capillaries. The lesion blanches on pressure and regains its color when the pressure removed. As the affected individual grows, the bleeding episode tends to increase in frequency and intensity.
- *Crushed spider appearance*—the typical lesion is cherry-red to purplish macule or small papule (**Fig. 37-19**) that resembles a *crushed spider*.
- *Epistaxis*—they often give rise to profuse hemorrhage, such as episodes of epistaxis. Severe bleeding may also occur from the gastrointestinal tract.



Fig. 37-19: Cherry red lesion seen on lower chin area in hereditary hemorrhagic telangiectasia.

Oral Manifestations

- *Site*—oral cavity, lips and tongue are most commonly affected.

- *Symptoms*—Severe oral hemorrhage may be experienced several times a day for weeks. At times there is gush of blood when involved areas are simply touched with cotton.
- *Appearance*—cherry red, often slightly raised, pinpoint or slightly larger lesions resembling a crushed spider is seen at any intraoral site, especially at the lip.
- *Hemorrhage during treatment*—hemorrhage can be encountered during dental treatment which encroach the affected area.

Diagnosis

- *Clinical diagnosis*—cherry red lesion seen on mucosal site may suspect this disease.
- *Laboratory diagnosis*—intrinsic defect in the endothelial cells permitting their detachment or defect in the perivascular supportive tissue bed, which weakens the vessels.

Management

- *Septal dermoplasty*—it is done in patient who has got repeated attacks of epistaxis. In this involve mucosa is removed and replaced by skin graft.
- *Pressure pack*—spontaneous hemorrhages may be controlled by pressure packs, particularly nasal bleeding.
- *Sclerosing agents*—sclerosing agent such as sodium morrhuate or sodium tetradecyl sulfate injected into the lesion is useful.
- *Electrocautery*—electrocautery is also useful, prophylactically in a lesion that is likely to cause bleeding.
- *LASER ablation*—it is also useful in some cases.
- *Other therapy*—other therapy like progesterone, estrogen, iron replacement therapy and occasionally blood transfusion is also indicated in severe patient.

Rare Hemorrhagic Disorders

- *Hypoprothrombinemia*—it is seen not only as a result of prothrombin deficiency but also due to factor V, VII, X and fibrinogen deficiency and when circulating inhibitors are present. The symptoms are similar to those of mild hemophilia. The amount of bleeding is variable and is usually brought about by trauma.
- *Factor XI deficiency or hemophilia C*—it is rare and regarded as an autosomal dominant trait. The bleeding tendency is erratic, but there is always the possibility of severe bleeding after minor surgical procedures, even though the patient may undergo numerous previous surgical procedures uneventfully.
- *Fibrin stabilizing factor deficiency*—it appears to be transmitted as autosomal recessive trait with high incidence of consanguinity. In the absence of this factor,

there are permanent peptide bonds between fibrin molecules so that fibrinogen monomer aggregates break up under certain conditions. Patients with this deficiency have severe post-surgical bleeding episodes which are typically delayed from 24 to 36 hours, hemoarthrosis and defective wound healing. Bleeding and clotting time are normal. Treatment is given prior to surgery by administration of factor XIII in small amounts.

- *Disseminated intravascular coagulation (DIC)*—it is an acquired bleeding disorder which is generally acute but may be chronic in onset in certain instances. The acute DIC is clinically severe with depletion of multiple clotting factors like fibrinogen, prothrombin, factor V and factor VIII and platelets are also reduced in number.
- *Hemangiomata*—these are congenital malformations and lesions are larger than hereditary hemorrhagic telangiectasia and tend to decrease in size at puberty. Larger malformation may persist through adult life. The lesion may vary in size, being unobstructive as macules or as predominant as pedunculated lesion. The macular lesions are less prone to hemorrhage. The macular lesion may occur anywhere in the oral cavity like on lip, gingiva, tongue, buccal mucosa or within the jaw bones. There is presence of mobile teeth, bleeding from the gingival crevice and expansion of the cortex of jaw bones in areas of involvement. Therapy can entail the use of Sclerosing agents such as sodium tetradecyl or morrhuate sodium injected into the lesion.
- *Hemorrhage of iatrogenic cause*—it may occur due to anti-coagulation therapy which is given in acute myocardial infarction, cerebrovascular accident, pulmonary embolism. Two types of anti-coagulants are used: heparin and the bishydroxycoumarin derivatives, which are used much more commonly than heparin in ambulatory patients. If patients are taking heparin, topical thrombin can be used to control hemorrhage due to dental treatment. If necessary, the heparin dose can be regulated under the able guidance of patient's physician. In an emergency, profuse hemorrhage can be controlled by factor IV protamine. If patients are taking bishydroxycoumarin, topical thrombin can be used to control hemorrhage. The action of bishydroxycoumarin derivatives may be reversed by giving intramuscular vitamin K1 but it takes about 12 to 24 hours for this reversal. In an emergency situation, patients are given plasma transfusion to restore the missing factors.

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Water soluble

- Vitamin B complex
 - Vitamin B₁ (thiamine)
 - Vitamin B₂ (riboflavin)
 - Vitamin B₃ (niacin)
 - Vitamin B₅ (pantothenic acid)
 - Vitamin B₆ (pyridoxine)
 - Vitamin B₈ (biotin)
 - Vitamin B₉ (folic acid)
 - Vitamin B₁₂ (cyanocobalamin)
- Vitamin C (ascorbic acid)
- Choline
- Inositol

Fat-soluble

- Vitamin A
- Vitamin D
- Vitamin E
- Vitamin K

a precursor steroid, niacin from tryptophan which is an essential amino acid, vitamin K and biotin by intestinal microflora.

Deficiency of vitamins may be primarily due to vitamin deficient diet or secondarily because of disturbances in intestinal absorption, transport in blood, tissue storage or metabolic conversion.

Causes of Vitamin Deficiency

- *Decreased intake*—decreased amount of intake of essential nutrients.
- *Impaired absorption*—impaired absorption from the alimentary tract.
- *Increased metabolism*—increased metabolism due to rapid growth.
- *Inadequate storage*—inadequate storage, fever and pregnancy.

Introduction

Vitamins are organic substances in food which are required in small amounts but which can not be synthesized in adequate quantities in the body and which are soluble in either fat or water. Vitamins are needed in small quantities to act as a cofactor in a variety of metabolic reactions. Your body needs only small amounts of vitamins. But because what the body manufacture is often not enough, these must be obtained from your diet and from supplements.

Vitamins occur in a natural and in a physiologically inactive form and are called as *provitamins*. They become activated only after conversion within the animals. For example, vitamin A exists in plants in the form carotene, which is activated in the liver.

Small amounts of vitamins can be synthesized endogenously. For example vitamin D is synthesized from

Water Soluble Vitamins

Water-soluble vitamins are found in yeast, grain, rice, vegetables, fish and meat. They are essential co-enzymes required in energy releasing mechanisms and in hemopoiesis. They also act as co-enzymes for metabolism of proteins, carbohydrates and fats.

B-complex Vitamins

Most of B-complex occurs in nature in the bound form within the cells of vegetables or animal tissues. The digestion for the liberation of vitamins and its absorption is a result of breakdown of cellular structures in the gut. Vitamin B-complex is not stored in appreciable amounts in the body tissues except vitamin B₁₂. Excretion of vitamins occurs in the kidney.

The oral signs of deficiency of vitamin-B occur in the oral tissues like tongue, mucous membrane and gingiva. It may result in dermatitis, stomatitis and gastritis and blood and bone marrow disorders. Degenerative changes of brain and nerves are also a characteristic feature of deficiency since nerve tissue depends on glucose. In hemopoiesis, vitamin B₁₂ and folate are essential for maturation of red cell precursors.

Thiamine (Vitamin B₁)

It is a vitamin for calm nerves. It is also known as 'aneurin'. It was discovered by Eijkman in 1897. It is a colorless basic organic compound composed of a sulfated pyrimidine ring.

Absorption and excretion

It is readily absorbed from both small and large intestines. The capacity of the human intestine to absorb this vitamin is limited to about 5 mg per day. It is phosphorylated by the liver and kidneys. In tissues, it is found as thiamin pyrophosphate.

Thiamin pyrophosphate is a coenzyme for decarboxylation of pyruvate to acetyl coenzyme A. Any excess supply of thiamine is excreted in the urine.

Sources (Fig. 38-1)

- *Cereals*—wheat germ, wheat flour and rice bran.
- *Pulses*—Soya beans, split Bengal gram, lentil, moth, dry beans and split red gram.
- *Vegetables*—lotus stems, capsicum and turnip greens.
- *Nuts and oilseeds*—groundnuts, pistachio nuts and mustard seeds.
- *Fruits*—apricots, pineapple and bael fruits.
- *Meat*—whole meat, pork and liver sheep.
- *Milk and milk products*—skimmed milk powder, cow's milk and khoa.

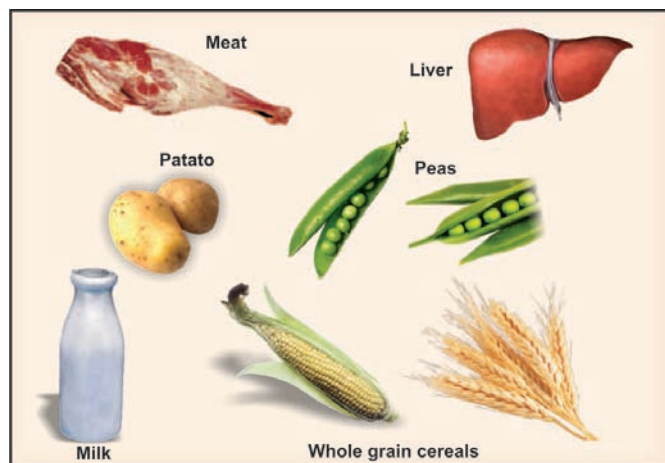


Fig. 38-1: Sources of vitamin B₁.

Daily requirements

- *Men*—1.3 mg daily
- *Women*—1.0 mg daily
- *Children*—1.1 mg daily
- *Pregnancy and lactation*—2 mg daily

Functions in the body

- *Growth*—it promotes growth, protects heart muscle and stimulates brain action.
- *Nervous system*—it plays an important role in the normal functioning of the entire nervous system.
- *Digestion*—it aids in digestion especially that of carbohydrates.
- *Diuretic*—it is a mild diuretic and it increases urine formation.
- *GIT*—it improves peristalsis and helps prevent constipation.
- *Blood cells*—it maintains the normal blood count and improves circulation.
- *Others*—it also reduces fatigue, increases stamina, prevent premature aging and senility by increasing mental alertness and promotes a healthy skin.

Deficiency symptoms

- *Nervous disorders*—when cells cannot metabolize the glucose aerobically, it affects the nervous system first since it depends entirely on glucose for its energy requirements. There is mental depression, nervous exhaustion and insomnia.
- *Digestive symptoms*—it occurs due to defective hydrochloric acid production in the stomach. Patient complains of loss of appetite, poor digestion, chronic constipation and loss of weight.
- *Heart*—there is accumulation of pyruvic acid and lactic acid derived from it, which produces vasodilatation and increases cardiac output. The heart muscle becomes lazy and fatigued and the auricles or the upper chambers of the heart lose their strength and it gradually enlarges. It may lead to a condition known as 'hypertrophy of the heart'.
- *Beriberi*—prolonged gross deficiency can cause beriberi. There are three types of beriberi-
 - Wet beriberi
 - Dry beriberi
 - Infantile beriberi
- *Others*—other diseases, which can be associated with it are
 - Wernicke's encephalopathy
 - Peripheral neuritis
 - Korsakoff's psychosis

Beriberi

It is marked by cardiac dilation with four chamber enlargement, pallor and flabbiness of myocardium.

Etiology

- **Diet**—it is caused due to eating polished rice in which outer husks is removed which contain thiamine.
- **Alcoholics**—it is commonly seen in chronic alcoholics due to their poor nutrition in general and also because alcohol interferes with intestinal absorption of thiamine.
- **Others**—it is often precipitated by infection, pregnancy and lactation.

Pathogenesis

- Deficiency of thiamine →incomplete metabolism of glucose →accumulation of pyruvic acid and lactic acid in tissue and body fluid →dilation of peripheral blood vessels →fluid may leak out through capillaries, producing edema →high cardiac output, heart dilation.

Clinical Features**Wet beriberi**

- **Symptoms**—pain in legs after walking due to accumulation of lactic acid.
- **Cardiac signs**—there is tachycardia and increased blood pressure, cardiomegaly, increased JVP and palpitations. There is also presence of sinus tachycardia and inverted T waves.
- **Skin**—skin is warm due to vasodilation.
- **Others**—edema may develop rapidly to involve leg, face and trunk.

Dry beriberi

- **Peripheral neuropathy**—peripheral neuropathy is a common feature of this disease.
- **Wasting of muscle**—in long standing cases, there is degeneration and demyelination of both sensory and motor nerve fibers resulting in severe wasting of muscles.

Oral manifestations

- **Hypertensive mucosa**—there is hypersensitivity of oral mucosa.
- **Symptoms**—pain in the tongue, teeth, jaws and face.

Management

- **Complete rest**—patient should advice the complete rest.
- **Thiamine**—thiamine 50 mg IM for 3 days then 10 mg 3 times daily by oral route.
- **Management of infantile beriberi**—infantile beriberi is treated via mother's milk. The mothers should receive 10,000 mcg twice daily. In addition, infants should be given thiamine in doses of 10,000 to 20,000 mcg IM once in a day for 3 days.
- **Precaution while giving thiamine to patient**—As such there are no toxic effects of thiamine. Any excess is excreted in the urine and not stored in any degree in the tissues or organs. Rare symptoms of overdose are tremors, herpes, edema, nervousness, rapid heartbeats and allergies. In rare cases, excessive supply of this vitamin may also adversely affect thyroid and insulin production.

Other deficiency symptoms

- **Wernicke's encephalopathy**—it is commonly seen in alcoholics with persistent vomiting. There is a classical triad of ocular abnormalities, ataxia and confusion. There are facial symmetrical areas of grayish discoloration. There is also bilateral symmetrical ophthalmoplegia and ataxia. Injection of thiamine should be given. 50 mg by slow intravenous injection followed by 50 mg daily by oral route for a week.
- **Korsakoff's psychosis**—in it, there is a predominant abnormality in mental function which is memory defect. There is profound impairment of memory recall and new learning ability.

Riboflavin (Vitamin B₂)

It is also called as the 'beauty vitamin'. It is a yellowish green fluorescent compound soluble in water. The word riboflavin is derived from two sources ribose, referring to ribose sugar found in several vitamins and enzymes and flavin meaning yellow. It is an essential component of coenzyme flavin mononucleotide and flavin adenine dinucleotide, involved mainly in a wide variety of oxidation-reduction reactions. It is stable to boiling in an acidic solution. It is decomposed by heat. It is also destroyed by ultraviolet light.

Absorption and excretion

It is readily absorbed from the intestinal tract and is phosphorylated in the wall of the intestine. It carried to the tissue of the body and incorporated into the cells enzymes. It is stored in liver, kidneys and heart. Riboflavin is excreted primarily in the urine and bile and sweat are other minor routes of excretion.

Sources (Fig. 38-2)

- **Cereals**—wheat germ, rice bran, bajara and barley.

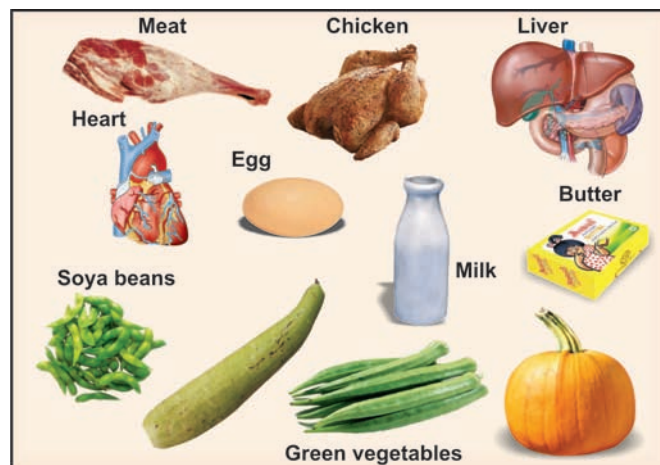


Fig. 38-2: Sources of vitamin B₂.

- *Pulses and legumes*—Soya beans, red gram, split green gram and peas.
- *Vegetables*—lotus stems, turnip greens, spinach, cauliflower and brinjals.
- *Nuts and oilseeds*—almonds, walnuts, chilgozas and mustard seeds.
- *Fruits*—papayas, raisins, custard apples and jack fruits.
- *Milk and milk products*—skimmed milk powder, cow's milk and whole milk powder.
- *Animals*—liver of sheep, eggs, mutton and prawn.

Daily requirement

- *For infants*—60 mcg value per kg of body weight daily.
- *Men*—1.5 mg daily
- *Women*—1.2 mg daily
- *Children*—1.3 mg daily
- *Pregnancy and lactation*—2 to 2.3 mg daily

Function in the body

- *Growth*—it is essential for growth and general health.
- *Metabolism*—it is involved in the metabolism of carbohydrates, fats and proteins. It is essential for normal tissue maintenance.
- *Nervous system*—it helps in functioning of the nervous system.
- *Digestion*—it helps in digestion and prevents constipation.
- *Eyes*—it alleviates eye strain and it is helpful in counteracting the tendency toward glaucoma.
- *Others*—it promotes a healthy skin, nails and hair and strengthens the mucous lining of the mouth, lips and tongue.

Causes of deficiency

- *Primary deficiency*—it occurs due to inadequate diet and also inadequacy of other essential nutrients including vitamins and proteins.
- *Secondary deficiency*
 - It may occur due to diseases of the intestinal tract.
 - Prolong use of psychological drug that interfere with production of flavin monophosphate.
 - Chronic alcoholism, burns and trauma.

Deficiency symptoms

- *Seborrheic dermatitis*—it affects the nasolabial fold and ala of the nose which exhibits a scaly gray dermatitis and consists of enlarged follicles around the side of the nose which is plugged with dry sebaceous material.
- *Ocular changes*—it consists of corneal vasodilatation, photophobia and superficial and interstitial keratitis. There may be itching and burning of the eyes.
- *Skin and nails*—it may also result in dull or oily hair, an oily skin, premature wrinkles on the face and arms and split nails.

- *Others*—malfunctioning of adrenal glands, anemia, vaginal itching and cataract.

Oral manifestations

- *Glossitis*—glossitis which begins with soreness of lateral margins of the tongue. Filiform papillae become atrophic while fungiform papillae remain normal or become engorged and mushroom shaped giving the tongue a reddened coarsely granular appearance. In severe cases, the tongue becomes glazed and smooth due to complete atrophy of the papillae and exhibits a magenta color.
- *Cheilitis*—lips become red and shiny because of desquamation of epithelium. Paleness of lips and cheilitis which is seen as maceration and fissuring at the angle of the mouth (**Fig. 38-3**). There is maceration at angle of mouth with pain on the opening mouth; it again results in fissuring and cracking with ulceration. As the disease progresses, angular cheilitis spread to the cheek, the tissues bleed easily and are painful if secondarily infected.



Fig. 38-3: Angular cheilitis seen in patient with riboflavin deficiency.

Management

- Riboflavin 25,000 to 50,000 mcg is given daily in divided doses.

Niacin (vitamin B₃)

It is also known as 'nicotinic acid'. Niacin is required for the formation of coenzyme NAD and NADP, which are important pyridine nucleotides which play an important role in redox reactions involving carbohydrate, protein and lipid metabolism. Deficiency of niacin leads to a disease called as 'pellagra' which means rough skin.

Absorption

It is absorbed from both stomach and intestine and stored in all tissues. It is excreted in the urine mostly as salts and to a smaller extent as free niacin.

Sources

- *Cereals*—rice bran, barley, wheat flour, jowar, wheat germ and finger millet.
- *Pulses and legumes*—peas, soya beans, red gram and split kesari.
- *Vegetables*—turnip greens, beet green, carrot leaves and potatoes.
- *Nuts and oilseeds*—groundnuts, sunflower, almonds and mustard seeds.
- *Fruits*—apricots, passion fruit, custard apple and bael fruit.
- *Animals*—liver of sheep, mutton, beef and prawns.
- *Milk and milk products*—skimmed milk powder, cow's milk and whole milk powder.
- *Tryptophan*—it can be endogenously prepared from tryptophan 60 mg of tryptophan gives 1 mg of niacin.

Daily requirements

- *Infants*—650 mcg per kg body weight daily.
- *Men*—17 mg daily.
- *Women*—13 mg daily.
- *Children*—15 mg daily.
- *Pregnancy and lactation*—12-15 mg daily.

Function in the body

- *Nervous system*—it is important for proper blood circulation and healthy functioning of the nervous system.
- *Gastrointestinal tract*—it is essential for the proper metabolism of proteins and carbohydrates.
- *Blood vessels*—it dilates the blood vessels and increases the flow of blood to the peripheral capillary system.
- *Hormone*—it is essential for the synthesis of sex hormone, estrogen, progesterone and testosterone as well as cortisone, thyroxine and insulin.
- *Others*—it helps to maintain a normal healthy skin.

Pellagra**Causes of deficiency**

- *Tryptophan deficiency*—if insufficient tryptophan is available for synthesis of niacin.
- *Diet*—dietary deficiency of niacin. High dietary levels of amino acid leucine antagonise the synthesis of NAD and NADP.
- *Miscellaneous*—chronic alcoholism, diarrhea and carcinoid syndrome.

Clinical Features

- *Prodromal symptoms*—it can be developed in 3 weeks with prodromal symptoms of loss of appetite, vague gastrointestinal disturbances and numbness or burning in various locations.
- *3 Ds of disease*—it is called as disease of 3-Ds
 - Dermatitis

- Diarrhea
- Dementia
- *Dermatitis*—there is an erythema resembling severe sunburns appears symmetrically over all parts of the body exposed to sunlight and especially on the neck. Affected area is well demarcated from the normal. In acute cases, skin lesions may produce vesiculation, cracking, exudation, crusting with ulceration and secondary infection. In chronic cases, dermatitis occurs as roughening and thickening of skin with brown pigmentation (**Fig. 38-4**).
- *Alimentary tract*—anorexia, nausea, dysphagia. Glossitis precedes the skin lesions. Non infective inflammation may extend through the gastrointestinal tract. Diarrhea is caused by atrophy of gastric epithelium followed by submucosal inflammation which is then followed by ulceration.
- *Nervous system*—delirium is the most common mental disturbance in the acute form and dementia in chronic cases.
- *Others*—there is also loss of appetite, irritability and burning sensation in different areas of the body.



Fig. 38-4: Thickening of skin with brown pigmentation seen in pellagra (Courtesy Dr Pincha).

Oral manifestations

- *Oral mucosa*—entire oral mucosa becomes fiery red and painful and salivation is profuse.
- *Glossitis*—the epithelium of the entire tongue is desquamated. The filiform papillae are most sensitive and disappear first; the fungiform papillae may become enlarged. The tongue becomes red swollen and beefy and in animals the deficiency leads to black tongue. In early stages, only the tip and margins of the tongue are swollen and red. In advanced cases, the tongue loses all the papillae and the reddening becomes intense. In this stage, the tongue becomes so swollen that indentation from the teeth are found along the borders of the tongue.

- *Stomatitis*—the mouth is sore and shows angular stomatitis, cheilitis. Tenderness, pain and ulceration begin at the interdental papillae and spreads rapidly.
- *Other features*—superimposed ANUG or Vincent's infection involving the gingiva, tongue and mucosa is common.

Management

- Niacin 10 mg or 10,000 mcg per day and vitamin B complex should also be given.
- Alcohol should be stopped.

Pantothenic Acid (vitamin B₅)

It is water soluble vitamin of the B complex group. It was discovered by Roger Williams in 1933. Tissue extracts from a variety of biological materials provided a growth factor for yeast. This growth factor is identified as pantothenic acid, derived from Greek word '*pantos*' meaning everywhere. It is a pale yellow oily liquid which is not crystallized, but its calcium crystallizes readily and this is the form in which it is generally available.

Absorption and excretion

It is not destroyed in neutral solution. It is liable to destruction by food, processing techniques, caffeine, sulphur drugs, sleeping pills and alcohol. It is absorbed from the alimentary tract and excreted in urine and mother's milk.

Sources

- *Cereals*—oatmeal, toasted wheat germ, brown rice and wheat flour.
- *Pulses and legumes*—Soya bean flour, split peas, lentil and blackeye peas.
- *Vegetables*—mushrooms, broccoli and cauliflower.
- *Nuts and oilseed*—peanuts, sunflower seeds and cashew nuts.
- *Meat*—calf liver.

Daily requirement

- *Men*—10 mg
- *Women*—10 mg
- *Children*—5.5 mg

Function in the body

- *Metabolism*—it is a part of enzyme system which plays a vital role in the metabolism of carbohydrates, fats and protein and in the synthesis of amino acids and fatty acids.
- *Formation porphyrins*—it is also essential for the formation of porphyrins, the pigment portion of the hemoglobin molecule.
- *Stimulation of gland*—it stimulates the adrenal glands and increases production of cortisone and other adrenal hormones.

- *Anti-stress factors*—it is primarily used as an anti-stress factor and protects against most physical and mental stress.
- *Recovery*—it increases vitality, wards off infections and speeds recovery from ill health.
- *Maintenance of normal growth*—it helps in maintaining the normal growth and development of the central nervous system.
- *Anti aging factor*—it prevents premature ageing and provides protection against any damage caused by excessive radiation.

Deficiency symptoms

- *Muscle tissue*—chronic fatigue, muscle cramps, painful and burning feet and muscular weakness.
- *Nervous system*—mental depression, irritability, dizziness and insomnia.
- *Gastrointestinal*—it may lead to gastric distress and constipation.
- *Others*—increase in tendency toward infection, graying and loss of hair, skin disorders, low blood sugar, low blood pressure and duodenal ulcer.

Management

- It is given in the dose of 1000 mg daily for 6 weeks.

Pyridoxine (Vitamin B₆)

It is an important coenzyme in the intermedullary metabolism of amino acids and complex glycolipids. It is a white crystalline substance soluble in water and alcohol.

Absorption and excretion

It is absorbed mainly in the jejunum and ileum of the small intestine by passive diffusion. It is widely distributed in various tissues and excreted mainly from the kidney. Small quantities of vitamins are excreted in the feces and in sweat.

Dietary sources (Fig. 38-5)

- *Cereals*—wheat germ, brown rice, wheat flour and barley.

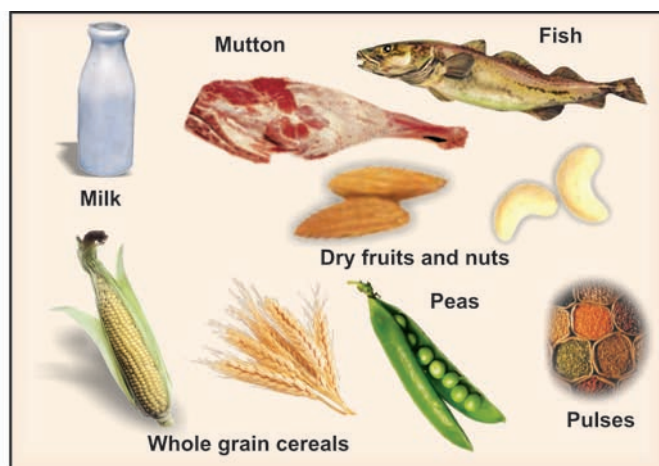


Fig. 38-5: Sources of vitamin B₆.

- *Pulses and legumes*—soya beans, lentil and lima beans.
- *Vegetables*—spinach, Brussels sprouts, potatoes and cauliflower.
- *Nuts and oilseeds*—sunflower seeds, walnuts and chestnuts.
- *Fruits*—bananas, avocados, prunes and raisins.

Daily requirements

- *Adults*—2 mg.
- *Children*—1.7 mg.
- *Infants*—0.1–0.4 mg.

Deficiency symptoms

- *Nervous*—peripheral neuropathy, mental retardation, irritability, mental confusion and nervousness.
- *Blood*—anemia, albuminuria and leukopenia.
- *Skin*—dermatitis and eczema.
- *Others*—kidney stones, inflammation of the colon, damage to the pancreas, loss of muscular control, migraine headache and premature senility.

Oral manifestation

- *Cheilosis*—cracking at the corner of the lip.
- *Glossitis*—inflammation of the tongue.
- *Others*—angular stomatitis, tooth decay and halitosis.

Management

- 10–50 mg daily in divided doses.

Biotin (vitamin B₇)

It functions as a coenzyme for four carbohydrates involved in fatty acid and amino acid metabolism. Previously was known as 'vitamin H'.

Daily requirements

- *Men and women*—100–200 mcg.
- *Children*—50–200 mcg.
- *Infants*—35 mcg.

Sources

- *Cereals*—rice bran, rice polishing, rice germ, barley, oatmeal and brown rice.
- *Pulses and legumes*—Soya bean flour, Soya bean, blackeye peas, spilt peas and lentil.
- *Vegetables*—mushrooms and cauliflower.
- *Nuts*—walnuts, peanuts and almonds.
- *Meat*—beef liver.

Function in the body

- *Metabolism*—it is involved in the metabolism of carbohydrates, proteins and fats.
- *Hair*—it is essential for the growth and health of the hair. It prevents premature graying of the hair as well as hair loss.
- *Others*—it helps to maintain the skin and nervous system in a sound condition. It controls proper distribution of color pigments.

Deficiency symptoms

- *Skin*—scaly dermatitis, eczema, seborrhea and prickling of the skin.
- *Hair*—it can cause alopecia and dandruff.
- *Nervous*—there is confusion, mental depression and drowsiness.
- *Muscle*—there is muscular weakness, extreme fatigue and lassitude.
- *Others*—anemia, lack of appetite, hearing abnormalities and lung infections.
- *Oral*—the fleshy part of the tongue may waste away.

Management

- 20 mcg of biotin is taken daily for 10 days IM can heal skin lesions. Oral biotin to be taken in amount of 400 mcg daily for 8–12 weeks. Shampoo coating 1% biotin can be useful in controlling excessive hair loss.

Folic Acid (vitamin B₉)

It is also known as 'folacin' or 'folate'. It is a water-soluble vitamin. It is a yellow crystalline substance sparingly soluble in water and soluble in acid solution. It undergoes fairly rapid destruction when heated in neutral or alkaline substances.

Absorption and excretion

It is absorbed along the entire length of the intestine, although the jejunum of the small intestine is the primary site for its absorption. About half of the folic acid stored in the body is in the liver. A small amount is excreted in the feces and urine but the additional amount is presumed to be metabolized and lost by cells coming off in the form of scales, from the body surface.

Daily requirements

- *Men and women*—100 mcg.
- *Children*—80 mcg
- *Infants*—25 mcg.
- *Pregnant women*—400 mcg.
- *Lactating women*—150 mcg.

Dietary sources

- *Cereals*—bajra, jowar, maize and wheat flour.
- *Pulses and legumes*—cowpeas, whole Bengal gram, split Bengal gram, split green gram, split black gram and lentil.
- *Vegetables*—spinach, cluster beans, ladies finger, curry leaves and French beans.
- *Nuts and oilseeds*—gingelly seeds, groundnuts and coconuts.
- *Meat and poultry*—eggs, liver of sheep and liver of goat.

Functions in the body

- *RBC*—folic acid in combination with vitamin B₁₂ is essential for the formation, maturation and multiplication of red blood cells.

- **Nerve**—it is necessary for the growth and division of all body cells, including nerve cells and for manufacturing a number of nerve transmitters.
- **Hair and skin**—it is essential for the health of skin and hair and helps to prevent premature graying of hair.
- **Pregnancy**—it is an important nutrient for the pregnant women and her developing fetus. Folic acid also improves lactation.
- **Others**—it helps in building of antibodies which prevent and heals infection. It also produces nucleic acids, RNA and DNA.

Deficiency causes

- **Decreased intake**—inadequate diet, impaired absorption, malabsorption states and intrinsic intestinal diseases.
- **Increased loss**—hemodialysis.
- **Increased requirement**—the body demands exceed the intake like in pregnancy, infancy, leukemia, hemolytic anemia.
- **Others**—impaired utilization, diseases of the upper small bowel where folate is mainly absorbed and idiopathic.

Clinical features

- **Anemia**—deficiency of folic acid cause anemia which often occurs in pregnant women and also children.
- **Skin**—loss of hair, grayish brown skin pigmentation can also occurs.
- **Reproductive disorders**—spontaneous abortions, difficulty during labor and high infant death can also occur. Loss of libido occurs in males.
- **Nervous**—dementia, mental depression and fatigue.

Oral manifestations

- **Atrophic glossitis**—filiform papillae disappear first and fungiform papillae remain prominent. In severe cases, fungiform papillae are lost and tongue becomes thick, smooth and fiery red (**Fig. 38-6**).



Fig. 38-6: Atrophic glossitis seen in folic acid deficiency patient.

- **Ulcerative stomatitis**—severe ulcerative stomatitis may be seen.
- **Other features**—swelling and redness of lips and lateral margin of the tongue.

Management

- A daily dose of 5,000 mcg to 10,000 mcg of folic acid is sufficient and a maintenance dose of 5000 mcg once in week is given in cases of megaloblastic anemia.

Cyanocobalamin (vitamin B₁₂)

Vitamin B₁₂ is a complex organomatrix compound called as cobalamine which is cobalt containing porphyrins. It is freely soluble in water. It is resistant to boiling in neutral solution, but it is liable to destruction in the presence of alkalis and acids.

Absorption and excretion

The presence of sufficient quantities of gastric juice is essential to facilitate its absorption in the intestine. Calcium and protein rich foods greatly help in the absorption of this vitamin from the intestine. It is stored in the liver which is capable of storing relatively large amounts of these nutrients. It is excreted in normal urine, stools and breast milk.

Sources (Fig. 38-7)

- **Animal origin**—it is unique amongst vitamins in that it is mostly found in foods of animal origin. Vegetarians are therefore advised to increase their intake of milk or take it in tablet form as a supplement.
- **Fish, meat and poultry**—sheep liver, goat liver, fresh shrimps, yolk eggs, goat meat, mutton, buffalo meat and whole eggs.

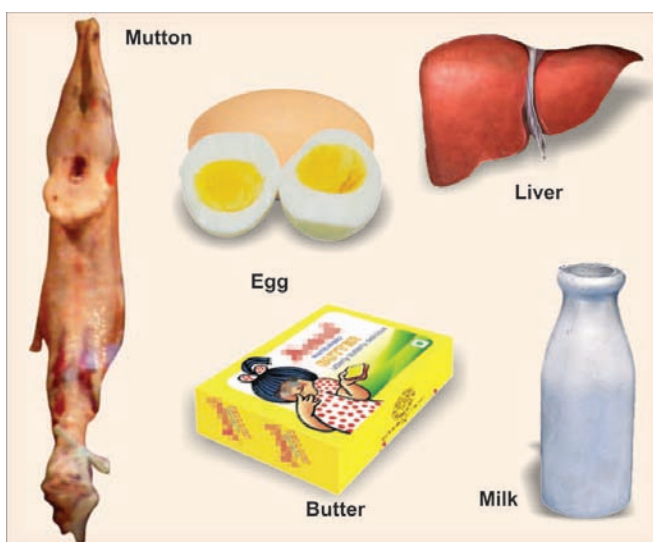


Fig. 38-7: Sources of vitamin B₁₂.

- *Milk and milk products*—skimmed milk powder, buffalo milk, cow's milk and curd.

Daily requirements

- *Men and women*—1 mcg.
- *Children*—0.2-1 mcg.
- *Infants*—0.2 mcg.

Functions in the body

- *Red blood cells*—like vitamin B₆ it is essential in the production and regeneration of red blood cells.
- *Nervous*—it improves concentration, memory and balance and relieves irritability.
- *Metabolism*—it is necessary for proper utilization of fats, carbohydrates and proteins for body building. It is also used in metabolism of folic acid.
- *Others*—it promotes growth and increases appetite in children.

Causes of deficiency

- *Congenital*—congenital deficiency without gastric atrophy.
- *Systemic diseases*—diseases of terminal ileum i.e. Crohn's disease.
- *Defective absorption*—there is defective absorption of vitamin B₁₂. There is chronic atrophic gastritis with failure of production of intrinsic factor.
- *Smoking*—studies show that smokers have lower levels of vitamin B₁₂ and folic acid than non-smoker.
- *Others*—inadequate diet and intrinsic factor deficiency.

Deficiency symptoms

- *Megaloblastic anemia*—deficiency of vitamin B₁₂ leads to megaloblastic anemia or pernicious anemia. However, pernicious anemia is a result of deficiency of intrinsic factor, which is essential for absorption of vitamin B₁₂ and hence a deficiency of vitamin B₁₂.
- *Age and sex*—it occurs in 5th to 8th decades of life. It is more common in men than in women.
- *Symptoms*—there is generalized weakness, numbness and tingling of the extremities. Fatigue, headache, dizziness, nausea, vomiting, diarrhea, loss of appetite, pallor and abdominal pain.

Oral manifestations

- *Tongue*—there is sore painful tongue, glossitis and glossodynia. Tongue is inflamed and is beefy red in color. Painful and burning lingual sensation. Small shallow ulcers resembling aphthous ulcers on the tongue with atrophy of papillae with a loss of normal muscle tone is called as 'hunter's glossitis'. Fiery red appearance of tongue due to inflammation and burning sensation.
- *Denture wearing discomfort*—discomfort in wearing dentures is due to weakened muscular tone.

Management

- *Oral*—it is used in a dose ranging from 6 to 150 mcg, helps in the treatment of lack of concentration, fatigue depression, insomnia, anorexia, poor memory and loss of weight.
- *Parenteral*—1000 mcg of vitamin given twice weekly in cases of anemia.

Vitamin C

It is also called as 'ascorbic acid' and 'antibiotic vitamin'. It is a modified simple sugar. It is the most active reducing agent. Its highest concentration is in the pituitary, adenoid, eye and WBCs. Stress and corticotrophin leads to loss of ascorbic acid from the adrenal cortex. It is a powerful antioxidant. It is necessary for normal maintenance of intercellular substances of connective tissue in bone and other tissue of mesenchymal origin.

Absorption and excretion

Absorption of ascorbic acid into the bloodstream takes place in the upper part of the small intestine. It is excreted by the kidney through the urine.

Dietary sources (Fig. 38-8)

- *Cereals, pulses and legumes*—red gram, peas, maize and Bengal gram.
- *Vegetable*—parsley, drumstick leaves, turnip greens, cabbage, bitter gourd, radish leaves, carrot leaves, beet greens cauliflower, cluster beans, tomatoes, spinach and ladies finger.
- *Nuts*—coconut dry and coconut milk.
- *Fruits*—Indian gooseberries, guavas, orange juice, limes, papayas strawberries, lemons, pineapples, custard apple, raspberry and mangoes.
- *Fish and meat*—Indian shad, Rohu, sheep liver and tenger fish.

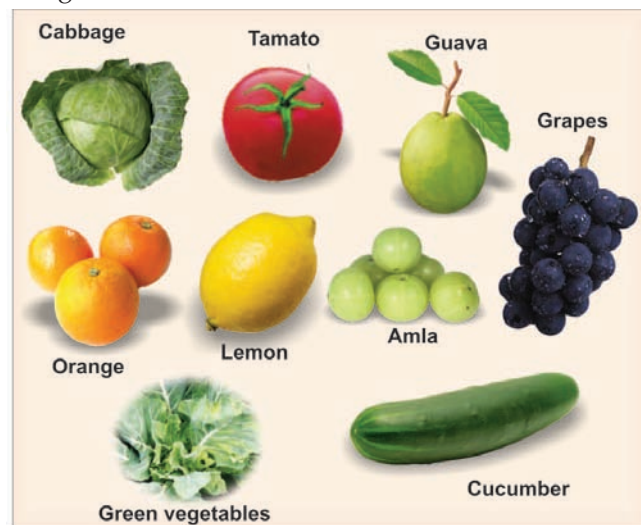


Fig. 38-8: Sources of vitamin C.

- *Milk and milk product*—khoa and skimmed milk powder and wholemilk powder.

Daily requirements

- *Men and women*—40 mg.
- *Infants*—25 mg.
- *Children*—40 mg.
- *Pregnant and lactating women*—80 mg.

Functions of vitamin C

- *Synthesis*—it is important in the formation of collagen, chondroitin sulfate and neurotransmitter.
- *Maintenance*—it is useful for maintenance of folate pool and mobility and phagocytic activity of neutrophils. It is also necessary for maintenance of bones and proper functioning of the adrenal and thyroid glands.
- *Absorption*—it enhances, the absorption of iron in the body.
- *Metabolism*—tryptophan, nor-epinephrine and tyrosine metabolism require vitamin C.
- *Others*—it promotes healing and protects against all forms of stress.

Deficiency symptoms

- *Mild deficiency*—it may appear in the form of lassitude fatigue, anorexia, muscular pain and greater susceptibility to infection.
- *Severe or prolonged deficiency*—a prolonged deficiency may cause scurvy.

Scurvy

Prolonged deficiency of vitamin C may result in scurvy. It is characterized by weakened blood vessels particularly microvessels having least muscular supports. There is also defective synthesis of osteoid which is derivative of collagen which results in impaired wound healing.

Pathogenesis

- *Defective collagen formation*—there is defective formation of collagen in connective tissues because of failure of hydroxylation of proline to hydroxyproline which is a characteristic amino acid of collagen.
- *Increase capillary permeability*—there is also increase in permeability of capillary (hemorrhage), anemia due to erythropoiesis and defective collagen formation.

Clinical features

- *Infantile scurvy*—lassitude, anorexia, painful limbs and enlargement of costochondral junction.
- *Folliculosis*—hair follicle rises above skin and there are perifollicular hemorrhages i.e. tiny points of bleeding occurring around the orifice of hair follicles with heaping of keratin like material.
- *Hemorrhage*—hemorrhage may occur in the joint, into nerve sheath under the nails or conjunctiva. Petechial hemorrhage occurs in buttocks, abdomen, legs, arms,

ankle and nail beds. There is also epistaxis, anemia and delayed wound healing.

- *Scorbutic child*—scorbutic child usually assumes a 'frog like' position and this may reflect as subperiosteal hemorrhage.
- *Edema*—edema of the limbs and face is a frequent finding in severe ascorbic acid deficiency.
- *Other features*—it may lead to premature aging, thyroid insufficiency and lower resistance to all infections.

Oral manifestations

- *Site*—it occurs chiefly in gingival and periodontal region.
- *Scurvy bud*—interdental and marginal gingiva is bright red, swollen, smooth, shiny surface producing an appearance known as 'scurvy bud' (Fig. 38-9). In fully developed scurvy, the gingiva becomes boggy, ulcerated and bleeds easily.
- *Color*—color changes to violaceous red.
- *Breath*—typical fetid breath of the patient with fusospirochetal stomatitis.
- *Severe cases*—in severe cases, hemorrhage and swelling of periodontal ligament membrane occurs followed by loss of bone and loosening of teeth which are exfoliated.



Fig. 38-9: Scurvy buds seen on gingiva in vitamin C deficiency patient

Diagnosis

- *Clinical diagnosis*—scurvy buds with increased hemorrhagic tendency may give clue to the diagnosis.
- *Laboratory diagnosis*—the calcified matrix material is not destroyed so that wide zone of calcified but nonossified matrix, called the 'scorbutic lattice' develops in the metaphysis. Anemia in scurvy is mild to moderate but may be severe.

Management

- Vitamin C 250 mg 3 times daily can be given.

Choline

It is a colorless crystalline compound which absorbs water quickly. It is highly soluble in water and alcohol. It is member of vitamin B group. It is present in foods as well as in the body in relatively large amount.

The body can make choline from methionine, an amino acid, with the aid of vitamin B₁₂ and folic acid.

Absorption and Excretion

It is highly soluble in water and alcohol. It is absorbed from the intestine and excreted mostly through the urine.

Sources

- *Fish and sea food*—it is available in liberal quantities in fish and sea foods.

Functions

- *Fat transportation*—it helps in the transportation of fats in the body and prevents accumulation of fat in the liver.
- *Nerve cell formation*—in combination with fatty acid and phosphorus, it stimulates the formation of lecithin, an important constituent of nerve cells in the body.
- *Memory*—it goes directly into the brain cells to produce a chemical that aids memory.

Deficiency Symptoms

- It may cause cirrhosis and fatty degeneration of the liver, high blood pressure and atherosclerosis.

Management

- *Doses*—it can be given in doses of 1000-2000 mg daily in divided doses.

Inositol

It is a crystalline compound which has sweet taste. It is highly soluble in water and is not destroyed by heat in neutral, acid and alkaline media.

- *Functions in the body*
 - *Fat transportation*—it is essential for transportation of fat in the body.
 - *Brain cells nourishment*—it is important in providing nourishment to the brain cells.
 - *Maintenance of cholesterol level*—it helps to lower cholesterol levels.
 - *Hair growth*—it also promotes the growth of healthy hair and helps to prevent its falling.
 - *Prevention of eczema*—it helps in preventing eczema.
- *Sources*—the most important sources is from liver, brewer's yeast, wheat germ, unrefined molasses, peanuts and cabbage.
- *Deficiency symptoms*—it can cause alopecia or patchy baldness, gastritis hypertension, fatty infiltration in the liver, hardening of the liver and eczema.
- *Doses*—it is given in the dose of 2 gm a day for 6-10 weeks.

Fat Soluble Vitamins

Common Properties

- They are soluble in fat.
- Bile salts are essential for their absorption.
- They are generally stored in the liver.
- They are not excreted in urine.

Vitamin A (Retinol)

Carotene is a yellow pigment found in vegetable foods. It is converted into vitamin A in the body. Vitamin A or retinol is found in foods of animal origin, while carotene is provided by foods of both plant and animal origin. Vitamin A is stored in liver. As a concentrated solution retinol is light yellow in color. It solidifies when cooled and has a mild pleasant odor.

Absorption

Approximately 80% of vitamin A is absorbed in the human system. It is passed along with fat through the lymphatic system in to the bloodstream. Absorption of vitamin A is increases if it is taken with fats. Absorption more rapid in men than in women. Absorption is of this vitamin is poor in cases of diarrhea, jaundice and abdominal disorders. Vitamin A which is not absorbed is excreted within one or two days in feces.

Sources (Fig. 38-10)

- *Plant sources*
 - *Cereals and pulses*—red gram, soya beans, bajra and lentil.
 - *Vegetables*—carrots, green leafy vegetables and spinach.
 - *Fruits*—sweet potatoes, papayas, tomatoes, mangoes, persimmon and raspberries.
- *Animal sources*—sheep liver, cow's milk, kidneys, fish liver oils.
- *Fats and edible oils*—butter, hydrogenated oil and ghee.

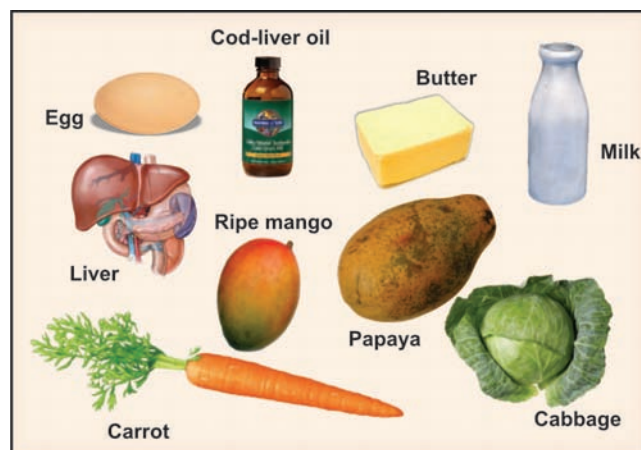


Fig. 38-10: Sources of vitamin A.

Requirements

- *Men and women*—600 mcg
- *Pregnancy and lactation*—950 mcg
- *Children*—600 mcg
- *Infants*—350 mcg

Functions in the Body

- *Epithelial tissue*—vitamin A helps in maintaining the integrity of epithelial tissue such as epithelial layer of skin, respiratory mucosa and esophagus and gastrourinary tract. Due to this, it builds up resistance to respiratory infection.
- *Structural integrity*—its function in the preservation of the structural integrity and normal permeability of the cell membrane as well as that of membrane of intracellular particles such as lysosomes and mitochondria.
- *Bone and teeth*—it accelerates the normal formation of bones and teeth.
- *Vision*—vitamin A also has a specific role on the physiological mechanism of vision.
- *Oxygenation*—it also increases permeability of blood capillaries thereby contributing to better tissue oxygenation.
- *Aging and senility*—it also prevents premature ageing and senility.
- *Synthesis*—it is required for synthesis of glucocorticoids and cholesterol.
- *Somatic growth*—vitamin A is required for somatic growth.

Deficiency Symptoms

- *Effect on growth*—failure of growth in young and collagenous tissue is affected.
- *Effect on eyes*—night blindness, dry conjunctiva, Bitot's spot, corneal xerosis, corneal ulceration or keratomalacia can occur. Xerophthalmia due to decrease in lacrimal secretion.
- *Keratinizing metaplasia*—the epithelial cells fail to differentiate. This means that the cells in the basal layer lose their specificity and tend to form a stratified squamous epithelium with keratin production. Keratinizing metaplasia of epithelial cells is usually evident in several organs such as bladder, vagina and skin and predisposes them to infection. Drying of skin and atrophy of sebaceous glands.
- *Effect on reproductive organs*—degeneration of germinal epithelium thus affecting reproduction causes sterility in males and cornification of vaginal epithelium in females.
- *Effects on bone*—imbalance between osteoblasts (bone forming cells) and osteoclasts (bone resorbing cells) causing aberrations in the shape of bone.

- *Skin disorders*—it may result in pimples, acne, boils and premature wrinkles.

Oral Manifestations

- *Teeth*—defective formation of enamel in teeth. Odontogenic epithelium fails to undergo normal histodifferentiation and morphodifferentiation, which results in increased rate of cell proliferation. Therefore epithelial invasion of pulpal tissue is characteristic of vitamin A deficiency. There is also distortion of shapes of the incisors and the molars.
- *Hypoplasia of teeth*—since the enamel forming cells are disturbed, enamel matrix is poorly defined so that calcification is disturbed and enamel hypoplasia results (Fig. 38-11).



Fig. 38-11: Enamel hypoplasia seen in patient with vitamin A deficiency.

- *Dentin*—dentin is too atypical in structure, lacking the normal tubular arrangement and containing vascular and cellular inclusions.
- *Caries*—there is increased caries susceptibility.
- *Eruption*—eruption is delayed. In prolonged deficiency, eruption ceases completely.
- *Alveolar bone*—alveolar bone is retarded in its rate of formation.
- *Gingiva*—gingival epithelium becomes hyperplastic; in prolonged deficiency it shows keratinization.
- *Periodontal disease*—tissue is easily invaded by bacteria that may cause periodontal disease and micro abscess formation.
- *Salivary gland*—major and minor salivary glands undergo typical keratinizing metaplasia.

Management

- Depending upon deficiency symptoms it is given in the dose of 7500-15,000 mcg per day for one month.

Hypervitaminosis A

- *Causes*—if more than 30,000 mcg of vitamin A is taken daily, it can produce toxic effect in adults if continued

for many months. In infants, toxic effect can be produced by the intake of more than 5,550 mcg per day.

- *Toxicity symptoms*—painful joints, thickening of long bones, anorexia, low grade fever, loss of hair, hepatomegaly, blurred vision, rashes, irregular menstruation, fatigue and headache.
- *Acute toxicity*—it results from a single massive dose and it manifests as abdominal pain, nausea, vomiting, headache, dizziness and sluggishness.
- *Chronic toxicity*—it may occur following ingestion of 12,000 mcg or more daily for prolonged periods. It is characterized by joint pain, hair loss, dryness and fissuring of lips, loss of appetite, low grade fever and weight loss.
- *Radiological findings*—radiographically, there is fragmentation of the distal fibular epiphysis and pronounced periosteal thickening.

Carotenemia

There is generalized yellowish skin and mucosa seen in patient. There is also excessive deposition of carotene which is result of high intake of foods containing carotene. Increased yellowishness is also seen in hyperlipidemia, diabetes, nephritis and hypothyroidism and in condition in which conversion of carotene to vitamin A is impaired by inborn metabolic error or hepatic disease.

Management

- *Restriction of diet*—there should be restriction of diet in the patients.

Vitamin D

It is also called as 'sunshine vitamin'. Vitamin D (1,25-dihydroxycholecalciferol) is one of the compound that are grouped together as the hydroxylated cholecalciferol. If vitamin D deficiency occurs in children and infants it is called as 'rickets' and if it occurs in adults it is called as 'osteomalacia'. Deficiency of vitamin D tends to cause hypocalcemia.

Forms

- D_3 —it is present in fish liver oils and animal fats. It is called as cholecalciferol.
- D_2 —it is obtained artificially by irradiation of ergosterol and called as ergocalciferol.

Absorption

Bile is essential for the absorption of vitamin D, fat helps in its absorption too. The vitamin is absorbed from the jejunum of the small intestine and is transported in the lymph chylomicrons to the bloodstream. Excretion of vitamin D

and its metabolites occurs primarily in the feces with the aid of bile salts.

Function in the Body

- *Maintenance*—the major function of vitamin D is the maintenance of normal plasma levels of calcium and phosphorous.
- *Teeth and bone*—it is very important for the proper formation of teeth and bones. It plays an important role in prevention of dental caries.
- *Thyroid gland*—it is necessary for the healthy functioning of parathyroid gland, which regulates the calcium levels in the blood.

Requirement

- *Infants and children*—0.01 mg.
- *Men and women*—0.01 mg.
- *Pregnancy and lactating women*—0.01 mg.

Sources (Fig. 38-12)

- *Fish and poultry*—cod liver oil, shark liver oil and eggs.
- *Fats and edible oils*—ghee and butter.
- *Sunlight*—it is the most important source the vitamin D.

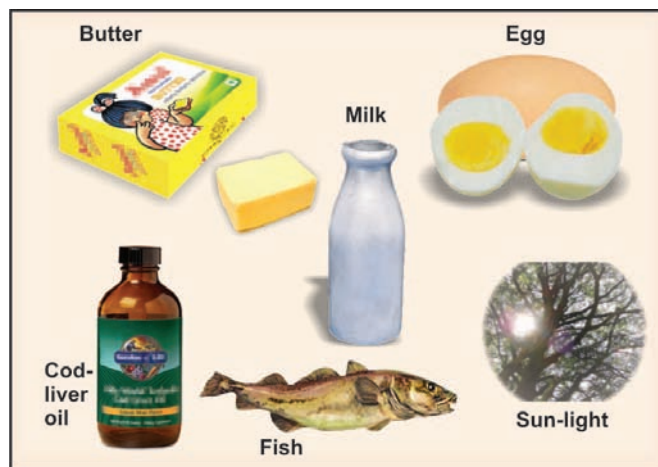


Fig. 38-12: Sources of vitamin D.

Pathogenesis

- *Overgrowth*—there is overgrowth of epiphyseal cartilage due to inadequate provisional calcification and failure of cartilage cells to form a matrix and disintegrates.
- *Formation of irregular masses*—there is persistence of distorted, irregular masses of cartilage many of which projects into the marrow cavity.
- *Deposition*—deposition of osteoid matrix on inadequately mineralized cartilaginous remnants.

- *Disruption*—disruption of the orderly replacement of cartilage by osteoid matrix with enlargement and lateral expansion of osteochondral junctions.
- *Microfracture*—there is abnormal growth of capillaries and fibroblasts in the disorganized zone because of microfracture and stresses on inadequately mineralized, weak, poorly formed bone.
- *Deformity*—deforming of skeleton due to loss of structural rigidity of the developing bone.

Vitamin D Deficient Rickets

The word 'Rickets' refers to any disorder in vitamin D calcium phosphorous axis which results in hypomineralized bone matrix that is failure of endochondral calcification. It develops in an area where sunlight is deficient. It results from inadequate extracellular level of calcium and inorganic phosphate, mineral necessary for new bone to calcify. Osteoid builds in excessive amounts because it fails to mineralize properly. Rickets occur in infants and children and osteomalacia common in adults.

Clinical features

- *Age*—it occurs in infants and children. In the first 6 month of life, tetany, convulsions are common manifestations due to hypocalcemia.
- *Sites*—the wrist and ankles are typically swollen. The changes in bone are found in the epiphyseal plates, metaphysis and the shaft.
- *Craniotabes*—localized area of thinning are sometimes present in the skull, so that a finger can produce indentation. This condition is called as 'craniotabes'. There is softening of posterior part of the parietal bone, which may be first sign of the disease.
- *Extremities*—patients has a short stature and deformed extremities. Children with rickets show bowing of legs.
- *Head*—excess of osteoid produces frontal bossing and squared appearance to the head.
- *Rachitic rosary*—deformation of chest results from over growth cartilage or osteoid tissue at the costochondrial junction producing 'rachitic rosary'.
- *Pigeon breast*—the weakened metaphyseal areas of the ribs are subject to pull of the respiratory muscles and thus bend inwards creating anterior protrusion of the sternum resulting in a pigeon breast deformity.
- *Harrison grooves*—the inward pull at the margins of diaphragm creates Harrison's grooves, girdling the thoracic cavity at the lower margin of the rib cage.
- *Lumbar lordosis*—the pelvis may be deformed. When an ambulatory child develops rickets, deformities are likely to affect the spine, pelvis and long bones causing 'lumbar lordosis'.

Oral manifestations

- *Teeth*—developmental abnormalities of dentine and enamel, delayed eruption and malalignment of teeth.
- *Caries*—there is higher caries index in rickets as compared to normal.
- *Enamel*—there may be hypoplasia of enamel; enamel may be mottled, yellow gray in color.
- *Pulp*—there are large pulp chamber, high pulp horns and delayed closure of root apices.
- *Malocclusion*—the osteoid is so soft that teeth are displaced leading to malocclusion of the teeth.

Radiographic features

- *Long bones*—the earliest and prominent manifestation is widening and fraying of epiphysis of the long bones. Bowing is a characteristic deformity seen in the weight bearing areas, fine trabeculae are reduced in number.
- *Green stick fractures*—it will be noted in many cases.
- *Jaw bone*—a thinning of jaw cortical structure such as the inferior mandibular canal, the lamina dura and the follicular walls of developing teeth has been described in rickets. The trabeculae become reduced in number. In severe cases, jaws appear completely radiolucent, so that teeth appear to be suspended in air.
- *Teeth*—if the disease occurs before 3 years of age, enamel hypoplasia is fairly common. The pulp cavities of deciduous teeth are grossly enlarged. The dentin is reduced to a thin margin separating the pulp cavity from enamel and cementum. The density of existing dentin appears to be normal and margins of pulp cavities are well and sharply defined. There is narrowing of the periodontal ligament space.

Diagnosis

- *Clinical diagnosis*—rachitic rosary, pigeon breast, Harrison grove with hypocalcification will diagnosed rickets.
- *Radiological features*—green stick fracture with narrowing of periodontal space.

Management

- *Dietary enrichment*—dietary enrichment of vitamin D in the form of milk.
- *Calcium gluconate*—if tetany is present, give calcium gluconate IV. Daily dose between 1000-2000 IU of vitamin D combined with 500-1000 mg of calcium. Curative treatment includes 2000 to 4000 IU of calcium daily for 6 to 12 weeks followed by a daily maintenance dose of 2000 to 4000 IU for a prolonged period.
- *Hormonal therapy*—hormonal therapy like flucytosine.

Vitamin D Resistant Rickets (Familial Hypophosphatemia, Refractory Rickets)

It is X-linked trait with some defect in reabsorption or metabolism.

Pathogenesis

- Vitamin D promotes the absorption of calcium and phosphorus from the intestinal tract, therefore diet deficient in vitamin D produces a negative calcium balance that in turn leads to under mineralization of the skeletal system. Thus rickets is caused by a lack of calcification of cartilage and osteoid tissue.

Causes

- *Decreased renal tubular reabsorption*—the disease is now recognized as a specific disorder characterized by hypophosphatemia associated with decreased renal tubular reabsorption of inorganic phosphates.
- *Genetic*—familial occurrence being inherited as X linked dominant trait.
- *Diminished calcium and phosphate absorption*—diminished intestinal calcium and phosphate absorption.

Clinical features

- *Age*—it is first recognized in children when they are about to walk.
- *Stature*—slight decrease in height of the patient and reduced growth and rickets like bone changes.
- *Signs and symptoms*—bowing of legs, enlarged epiphysis, bone pain, muscle weakness and vertebral fracture. Bony outgrowth at the site of muscular attachment and around joints may limit the movement

Oral manifestations

- *Dentin*—widespread formation of globular hypocalcified dentin with clefts and tubular defects occurring in the region of pulp horns. Gross reduction in amount and quantity of dentin which results in abnormally wide root canal and large pulp chambers with faulty calcification and marked interglobular space in dentin.
- *Pulp*—pulp horns are elongated and extend high often reaching the dentoenamel junction.
- *Periapical infection*—because of this defect there commonly is invasion of the pulp by microorganisms without demonstrable destruction of the tubular matrix. Thus there is often periapical involvement of a grossly normal appearing deciduous or permanent tooth followed by the development of multiple gingival fistulae. Tract is frequently present in dentinoenamel junction or even outer enamel surface. This tract remains patent and may result in early pulpal infection developing in abscess or carious lesion.
- *Cementum*—there is formation of abnormal cementum.

Radiographic features

- *Lamina dura*—lamina dura around the teeth is frequently absent or poorly defined. Osteoporotic bone with thinned dental crypts.
- *Jaw bones*—in some cases, only a relatively faint outline of the jaw is seen, with almost complete absence of

trabeculation. In such cases, there is no evidence of the normal cortical layer of bone around the follicle of developing teeth; as a result they appear unsupported by solid tissue.

- *Long bone*—it shows persistent deformities fracture and pseudo fractures.
- *Teeth*—thin enamel cap and enlarged pulp chambers and root canals. The dentin is reduced to a thin margin separating the pulp cavity from the enamel and cementum.
- *Infection*—high incidence of periapical and periodontal abscess which spread diffusely through the bone. The reasons for this is that as enlarged pulp forms in these teeth reaching nearly to dentinoenamel junction and there may be invasion of microorganisms in areas of hypoplastic enamel leading to pulpal necrosis. Alveolar pattern of bone is abnormal.

Diagnosis

- *Clinical diagnosis*—similar clinical feature of vitamin D dependent rickets with absence of teeth hypocalcification.
- *Radiological features*—multiple periapical infection with loss of lamina dura.
- *Laboratory diagnosis*—biopsy shows deposition of bone salts in cartilage matrix between the rows of hypertrophic cells so that these cells are not invaded and destroyed by capillaries. The histological feature is characterized by a broad zone between the multiplying cartilage cells and the shaft the so called 'rachitic metaphysis' which is composed of tongues of cartilage which extend down toward the shaft and are separated from one another by collection of capillaries.

Management

- *25-hydroxy cholecalciferol*—it is given lower doses is useful than conventional vitamin D.
- *Calcitrol and phosphate*—multiple daily doses of phosphate and calcitrol are given.
- *Endodontic therapy*—it is done tooth with periapical pathology.

Osteomalacia

It is also known as 'adult rickets' and only flat bones and diaphyses of long bones are affected. It is most commonly seen in post menopause females with a history of low dietary calcium intake and little exposure to UV light.

Clinical features

- *Age and sex*—it is seen in adults and pelvic deformities are commonly seen in females.
- *Bone*—remodeling of bone occurs in the absence of adequate calcium resulting in softening and distortion of the skeleton.

- *Symptoms*—the majority of patients has bone pain and muscle weakness of varying severity.
- *Others*—there is increased tendency towards fracture, peculiar waddling or penguin gait, tetany and green stick bone fractures.

Oral manifestations

- *Severe periodontitis*—there is incidence of severe periodontitis in some cases of osteomalacia.

Radiographic features

- *Pseudofracture*—a poorly calcified ribbon like zone extending into bone at approximately right angles to the periosteal margin. They are partial or complete fracture without displacement in which callus has been formed but there is no calcium available to be deposited, thus healing process is not complete and fracture remains apparent radiographically. It is also called as “Looser’s zone”. Osteoid tissue is formed in the defect but there is no calcium available to be deposited in the osteoid.
- *Jaws*—pseudofracture of the jaws near the angle has also been noted.
- *Bone*—individual bony trabeculae may be sparse and unusually coarse in intraoral periapical radiograph.
- *Lamina dura*—the lamina dura may be thin or absent in long standing and severe cases of osteomalacia.

Diagnosis

- *Clinical diagnosis*—bone pain, muscle weakness, fracture and waddling gait.
- *Radiological diagnosis*—pseudofracture can be seen radiologically.
- *Laboratory diagnosis*—there is elevation of serum alkaline phosphatase to three or more times its normal levels. Serum phosphorus is low due to increased phosphorus excretion in response to reduction of serum calcium. Serum calcium levels are usually on the lower side.

Management

- *Doses*—patients with osteomalacia due to intestinal malabsorption require a larger dose of vitamin D and calcium i.e. 40,000 to 1, 00,000 IU of vitamin D and 15 to 20 gm of calcium lactate per day.
- *Effect of hypervitaminosis D*—if patient is given in excess of vitamin D, patient may suffer from nausea, vomiting, constipation, drowsiness and sign of renal failure. There is also metastatic calcification in arteriole and kidneys and other tissues.

Vitamin E (Tocopherol)

It is also called as ‘anti-aging’ factor. The word tocopherol is derived from the word *tocos* meaning child birth and *pheros* meaning to bear. It is yellow, oily liquid freely soluble in fat solvents. Tocopherol alpha, beta, gamma, lambda

have been obtained from natural sources and their relationship with fertility and prevention of muscular dystrophy have been found. They are not destroyed by heat even at room temperature or above 100°C. They are destroyed by UV light.

Absorption and Excretion

Vitamin E in the diet is absorbed from the gastrointestinal tract by a mechanism similar to that for other fat soluble vitamin. It enters the bloodstream via the lymph. About 1/3rd of the vitamin is excreted in the bile and the balance is excreted in the urine.

Sources (Fig. 38-13)

- *Vegetables oils*—the richest sources of vitamin E are cold pressed crude vegetable oils, especially wheat germ, sunflower seeds, safflower and soya bean oils.
- *Cereals*—raw sprouted seeds and grains especially whole wheat are moderate sources.
- *Animal*—meats, eggs are minor sources of vitamin E.

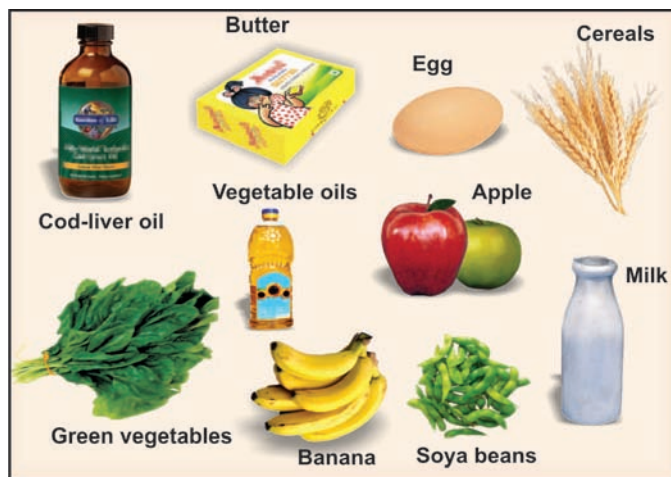


Fig. 38-13: Sources of vitamin E.

Daily Requirements

- *Men*—15 mg.
- *Women*—12 mg.
- *Children*—8.3 mg
- *Infants*—4-5 mg.

Functions in the Body

- *Reproductive function*—protective effect of vitamin E on reproduction and prevention of sterility. All the three layers of embryo ectoderm, mesoderm and endoderm are preserved by vitamin E.
- *Blood flow and clotting mechanism*—vitamin E dilates the capillary and enables the blood to flow freely into the

blood deficient muscle tissue thus strengthening both the tissue and the nerves supplying them. It also dissolves the blood clot and also prevents their formation.

- *Electron transport system*—it functions as a cofactor in the electron transport system.
- *Healing*—it prevents the formation of excessive scar tissue and in some instance, even melts away unwanted scar tissue.
- *Prevention*—it is required for prevention and storage of creatinine in muscles. It has ability to prevent hepatic necrosis in animals. Prevents vitamin A from destruction and helps in its storage in tissue.

Deficiency Symptoms

- *Reproductive*—abortion of fetus in females and atrophy of spermatogenic structures in males leading to permanent sterility.
- *Muscles*—it causes degenerative changes in muscles. There is muscle fiber atrophy which is replaced by connective tissue.
- *Heart*—there is necrosis and fibrosis of heart muscles.
- *Blood capillaries*—deficiency may lead to degenerative changes in the blood capillaries which in turn lead to heart and lung diseases, pulmonary embolism and brain stroke.

Oral Manifestations

- Loss of pigmentation, atrophic degenerative changes in enamel of vitamin E deficient rates.

Management

- Vitamin E is given in the doses of 100 -400 mg daily.

Vitamin K (Phylloquinone)

It is essential for the production of a type of protein called Prothrombin and other factors involve in the blood clotting mechanism. Hence it is known as '*anti-hemorrhagic vitamin*'. It is not easily destroyed by light, heat or exposure to air. It is destroyed by strong, acids, alkalis and oxidizing agents.

Forms

- K_1 —it is the form which occurs in plants.
- K_2 —it is produced by most bacteria present in human intestine if not supplied in the diet.

Requirements

- *Men and women*—70-140 mcg.
- *Children*—35-75 mcg.

Sources (Fig. 38-14)

- *Vegetables*—kale, turnip greens, spinach, broccoli, cabbage, lettuce, asparagus and watercress.

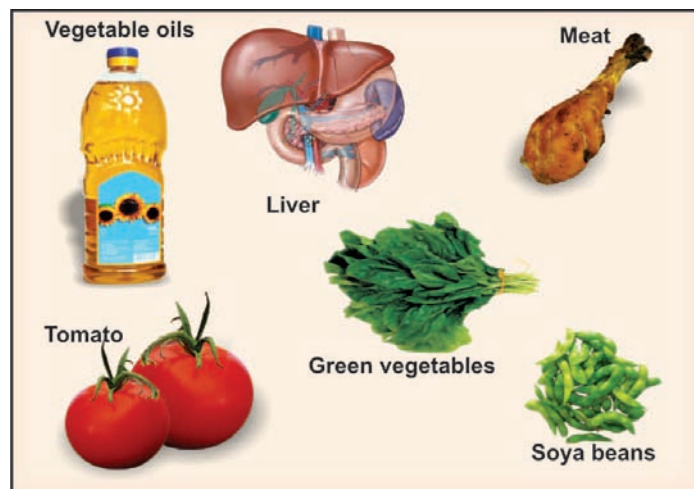


Fig. 38-14: Sources of vitamin K.

Functions in the Body

- *Synthesis*—it is essential for the hepatic synthesis of coagulation factors II, V, VII, IX and X.
- *Clotting*—it prevents hemorrhage only in cases when there is defective production of prothrombin.
- *Oxidative phosphorylation*—it acts as a cofactor in oxidative phosphorylation associated with lipid.

Effects of Deficiency

- Prolongation of clotting time and a tendency to bleed profusely.
- There may be nasal bleeding.

Oral Manifestations

- Gingival bleeding can also occur in cases of vitamin K deficiency.

Management

- It is given in dose of 10-20 mg daily.

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Introduction

Man is a complex biologic unit in a complex environment. Duncan defined metabolisms as sum of total tissue activity as considered in terms of physicochemical changes associated with and regulated by availability, utilization and disposal of protein, fat, carbohydrate, vitamins, minerals, water and influence which the endocrine exerts on these processes. Alternation from this normal metabolic process constitutes the disturbances of metabolism.

Disturbances in Protein Metabolism

All living tissues, whether plant or animal contains protein. Proteins constitute the most important group of foodstuffs. In addition to contributing to cells and intercellular materials, proteins and their constituent amino acids are important in the formation of hormone, enzymes, plasma proteins, antibodies and numerous other physiologically active substances. 1 gm of protein is required for each kg of body weight. It is required in increased quantity in later half of pregnancy, during lactation and infancy, childhood and adolescence.

Protein Energy Malnutrition

Protein energy malnutrition is characterized by energy deficit in macronutrient and micronutrient. Protein is the most important group of foodstuff. In addition to contributing to cells and intercellular material, proteins also help in the formation of hormones, enzymes, plasma protein antibodies and numerous physiologically important substances.

Types

- *Marasmus*—it is an overall deficit of food intake which results from near starvation with deficiency of protein and non-protein nutrient.
- *Kwashiorkor*—it is associated with primary dietary protein deficiency.
- *Marasmic kwashiorkor*—this is combined form of between the two extremes.

Etiology

- *Prolonged illness*—it occurs due to prolonged febrile illness, massive burns and large chronic ulcer.
- *Chronic infection*—chronic infections like urinary tract infection, tuberculosis and parasitic infection.
- *Endocrine disease*—hyperthyroidism and hyper-metabolic state which interfere with utilization.
- *Other*—stress, starvation and persistent vomiting and diarrhea.

Clinical Features

- *Age*—it occurs between the age of 1 and 3 years in children.
- *Marasmus*—there is emaciation due to weight loss, wasting of subcutaneous fat and muscles. Marasmus can also cause infantile gastroenteritis.
- *Kwashiorkor disease*—there is generalized edema, flaky paint dermatosis, thinning, and reddening of hair. There is also enlarged fatty liver. Kwashiorkor is distinguished from Marasmus by the presence of edema and the relatively less severe degree of body wasting.
- *Striped flag appearance*—alternating episodes of under nutrition and adequate nutrition may cause hair to have dramatic striped flag appearance. This is seen in kwashiorkor disease.

Oral Manifestations

- *Tongue*—bright reddening of tongue with loss of papillae. In kwashiorkor patient may have edema of tongue and may develop scalloping around the lateral margins due to indentation of the teeth. Papillary atrophy may be present and dorsum of the tongue assumes an erythematous and smooth appearance.
- *Perioral finding*—there is bilateral angular cheilosis, fissuring of lip. Loss of circumoral pigmentation.
- *Xerostomia*—mouth becomes dry. However, there is reduced caries activity due to lack of substrate carbohydrate.
- *Epithelium*—epithelium easily becomes detached from underlying tissue, leaving raw bleeding surface.
- *Jaws*—decreased overall growth of jaws, delayed eruption, retarded growth of incisors and molars.
- *Periodontal membrane*—The gingiva and periodontal ligament membrane exhibit varying degrees of degeneration.
- *Teeth*—deciduous teeth of children exhibits linear hypoplasia of the teeth.
- *Maxillofacial gangrene*—is severe infection that spread in mouth from necrotizing gingivitis.

Diagnosis

- *Clinical diagnosis*—striped flag appearance, weight loss, wasting of muscle, and flaky paint will give clue to diagnosis.
- *Laboratory diagnosis*—it may cause mild anemia which is normocytic and normochromic. The reticulocyte count is normal and the bone marrow tends to become hypocellular.

Management

- *Antibiotics*—infection should be treated with antibiotics.
- *Nutritional supplement*—it is usually supplied with milk based formulae. Full fat desiccated milk products can be fortified with corn oil and maltodextrin.

Amyloidosis

It is also called as 'amyloid disease'. It is deposition of amyloid in the tissue.

Forms of Amyloid

- *Type A* (secondary) amyloid is a fibrillar protein of unknown origin that is seen in prolonged inflammatory disease, genetic disease and syndromes such as familial Mediterranean fever.
- *Type B* (primary) amyloid is thought to be immunologic in origin because of its sequence homology with the NH₂

terminal end of immunoglobulin light chain. It is commonly seen in patient with multiple myeloma and macroglobulinemia.

- *Type C*—it includes amyloid of aging, localized nonspecific amyloid and pheochromocytomas.

Types

- *Primary amyloidosis*—there is no evidence of preceding or existing disease.
- *Amyloidosis associated with multiple myeloma*—it usually affects adult patients.
- *Secondary amyloidosis*—associated with variety of chronic inflammatory disease.
- *Localized amyloidosis*—it is characterized by small localized deposits of amyloid in the skin, bladder and respiratory tract.
- *Familial amyloidosis*—it is rare condition such as familial Mediterranean form or familial amyloidosis with polyneuropathy.
- *Hormone related amyloid*—it is associated with tumors of endocrine cells which secretes peptide hormones.

Etiology

- *Collagen diseases*—like rheumatoid arthritis.
- *Chronic infection*—chronic infections like tuberculosis, osteomyelitis, regional enteritis and ulcerative colitis.
- *Malignant disease*—malignant diseases like multiple myeloma, Hodgkin's lymphoma and renal cell carcinoma.

Clinical Features

- *Site*—commonly affected organs are kidneys, heart, G.I. tract, liver, respiratory tract, skin, eyes, adrenals, nerves and spleen. There may be primary localized collection of amyloid.
- *Symptoms*—the general symptoms are fatigue, weakness, ankle edema, dyspnea, paresthesia, orthostatic hypotension and weight loss.
- *Signs*—purpuric spots caused by hemorrhage resulting in formation of amyloid deposits in the blood vessels. Superficial waxy lesion occurs on the eyelids, nasolabial folds, neck, axilla or perineum and they may bleed on pressure.
- *Myocardium*—congestive cardiac failure is a common problem due to amyloid deposits in myocardium.
- *GIT involvement*—there is hepatomegaly, malabsorption or colitis may develop.

Oral Manifestations

- *Symptoms*—there are difficulties in chewing, swallowing or talking. The speech difficulty is due to paresis of the

vocal cord resulting from deposit of amyloid in the upper third of larynx.

- *Appearance*—tongue is enlarged and studded with small garnet colored enlargements along with nodes of cheeks and lips.
- *Signs*—mobility of the tongue is decreased. Yellowish nodules are present along the lateral border of the tongue and impressions from the teeth are also visible.
- *Skin lesion*—it may diffusely involve the face or may present as small elevated yellow nodules.
- *Macroglossia*—fibrous glycoproteins are deposited in submucosa as well as in deeper muscular layer of tongue. Amyloid deposition of tongue is found resulting in macroglossia of tongue. It is seen in both primary and secondary form.
- *Gingiva*—the gingiva may be infiltrated and may be bluish, spongy and hypertrophied.
- *Salivary gland involvement*—xerostomia may result from salivary gland involvement.

Diagnosis

- *Clinical diagnosis*—macroglossia with amyloid deposits can be seen on the face of patient.
- *Scintiscanning*—scintiscanning with ^{99m}Tc to localized soft tissue deposits.
- *Laboratory diagnosis*—congo red is used to diagnose amyloid which shows birefringence and dichroism.

Management

- *Alkylating agents*—you can treat amyloidosis by alkylating agents like melphalan.
- *Combination therapy*—combination therapy using melphalan, prednisone and fluoxymesterone has reported significant improvement.
- *Renal transplantation*—renal transplantation may be indicated. It will arrest the progression of bone lesion.
- *Debulking of lesion*—debulking of lesion can be carried out in case of macroglossia of tongue.

Porphyria

It is a group of disorders in which there is defect in enzymes involved in heme mechanism. These will result in accumulation of porphyrins.

Classification

- *Erythropoietic porphyria*—there is excessive abnormal porphyrins formation in developing erythrocytes. It is localized to bone marrow.
 - *Uroporphyrin*
 - *Protoporphyrin*.

- *Hepatic porphyria*—in it, liver is the site of excessive porphyrin formation.

- Acute intermittent porphyria
- Porphyria variegata
- Porphyria cutaneous tarda
- Hereditary coproporphyria.

Clinical Features

- *Sex*—it is transmitted as non-sex linked recessive character and both sexes are equally affected.
- *Urine*—the first sign is excretion of red urine containing uroporphyrin which may be noted at birth or apparent at first two year after birth.
- *Photosensitivity*—excessive deposition of the excess porphyrin in the skin lead to photosensitivity. It is absent in the neonatal period but may become apparent during first few years after it becomes exposed to sunlight.
- *Vesiculobullous lesion*—vesicular and bullous eruption appears on face, back and hand, i.e. exposed parts. Vesicle contains a serous fluid which exhibits red fluorescence. Ruptured vesicle heals slowly and leaves depressed pigmented scars.
- *Anemia*—there is often coexisting anemia.
- *Hepatic porphyria*—in this disease, there is abdominal crisis and psychological or metal symptoms. There is demyelination of nerve fibers.

Oral Manifestations

- *Teeth*—the porphyrin has an affinity for calcium phosphate and due to this, deposition of porphyrin occur in dentin. Deciduous and permanent teeth show red or brownish discoloration which under ultraviolet light exhibits red fluorescence due to incorporation of porphyrins during development.
- *Oral mucosa*—bullous, erosive lesions of oral mucosa may be present.
- *Cheilitis and periodontitis*—there is also atrophic cheilitis and advanced periodontal diseases.

Diagnosis

- *Clinical diagnosis*—red urine, photophobia and oral ulcerative lesion will give clue to diagnosis.
- *Laboratory diagnosis*—urine will demonstrate 5-aminolevulinic acid and porphobilinogen in the urine.

Management

- *Venesection*—this will reduce hepatic iron overload.
- *Stoppage of triggering drugs*—use of triggering drugs should be stopped immediately.
- *Intravenous drug*—intravenous drugs of hem arginate, fluids, electrolytes and glucose.

Disturbances in Lipid Metabolism

Lipids are a heterogeneous group of organic compounds which are relatively insoluble in water but, soluble in solvent such as ether, chloroform and benzene.

Classification

- *Simple lipids*—they are esters of fatty acids with various alcohols, i.e. natural fats and waxes.
- *Compound lipids*—they are esters of fatty acids containing groups other than and in addition to an alcohol and fatty acids.
- *Derived lipids*—they are derivatives obtained by hydrolysis of the simple and compound lipids, which still possess the general characteristics of lipids.
- *Miscellaneous*—it includes carotenoids, vitamin E and K.

Function of Lipids

- *Fuel*—lipids act as fuel in the body.
- *Insulation of body*—they insulate the body and protect various internal organs.
- *Building blocks*—they provide building blocks for different high molecular weight substance.
- *Essential fatty acids*—lipids supply the essential fatty acids which cannot be synthesized in the body and are essential in the diet for normal health and growth.
- *Proper functioning of nervous system*—the nervous system is particularly rich in lipids and is essential for proper functioning.
- *Vitamin maintenance*—some vitamins like A, D, E and K are fat soluble, hence lipid is necessary for these vitamins.
- *Carrier*—lipoprotein is also 'carrier' of triglycerides and cholesterol.

Classification of Disorders

- *Non-lipid reticuloendotheliosis*—it is also called as histiocytosis X, idiopathic histiocytosis and Langerhans cell disease. It is inflammatory reticuloendothelioma condition with evidence suggesting that it may be reaction to some type of infection. There is pathological accumulation of histiocytes and eosinophilic leukocytes. It is of 3 types. Nowadays, all these three diseases is called as *Langerhans cell histiocytosis (LCH)*:
 - Hand-Schüller-Christian disease.
 - Eosinophilic granuloma of bone (chronic localized Histiocytosis X)
 - Letterer Siwe disease (acute disseminated histiocytosis X).
- *Lipid reticuloendotheliosis*—it is disturbance in sphingo-myelin and glucosyl ceramide metabolism.
 - Gaucher's disease

- Niemann-Pick disease
- Tay-Sachs disease.

Langerhans Cell Histiocytosis

Nomenclature

In 1893, Hand reported child patient with diabetes insipidus, hepatosplenomegaly with punched out lesion of skull. In 1915 Schüller presented same features with another child patient. In 1920 Christian presented two cases with same features. So initially first entity called Hand-Schüller-Christian disease was established. Histologically, it is now classified as non-lipid reticuloendotheliosis.

In 1924, Letterer reported infant patient who has got low grade fever, hepatosplenomegaly, lymphadenopathy, and cutaneous petechiae with evidence of reticuloendothelial proliferation. In 1933 Siwe described similar cases. So term Letterer Siwe disease term was established.

In 1940, Otani and Ehrlich presented seven cases of solitary granuloma of bone which is subsequently called as eosinophilic granuloma of bone.

In 1953, Lichtenstein proposed a term Histiocytosis X for all the three diseases to indicate that basic disease process is of unknown etiology.

Nowadays, dominant histolytic cells identified as antigen processing Langerhans cells. So it is now called as Langerhans cell histiocytosis (LCH). All the three diseases are now called separated clinical entity under the heading of Langerhans cell histiocytosis.

Variant

- *Hand-Schüller-Christian disease*—it is also called as 'multifocal eosinophilic granuloma', 'chronic disseminated histiocytosis X' or 'xanthomatosis'. It is characterized by widespread skeletal and extraskelatal lesions and chronic clinical course. It is result of error in the metabolism of cholesterol and its esters.
- *Letterer Siwe disease*—It is an acute fulminating disease, which invariably occurs in infants usually before the age of 3 years.
- *Eosinophilic granuloma*—it is also called as 'unifocal eosinophilic granuloma'. It is the lesion of bone which is primarily histiocytes proliferation with an abundance of eosinophilic leukocytes by no intra- or extracellular lipid accumulation.

Clinical Features

Hand-Schüller-Christian disease

- *Age and sex*—it is more common in boys than girls 2:1. Occur in early life usually before age of five.
- *Classic triad*
 - Single or multiple areas of punched out bone destruction in skull.

- Unilateral or bilateral exophthalmos.
- Diabetes insipidus.
- *Facial asymmetry*—involvement of facial bone which is commonly associated with soft tissue swelling and tenderness causing facial asymmetry.
- *Signs*—otitis media and skin may sometimes exhibit papular or nodular lesion.
- *Progress*—course is chronic with numerous remissions and exacerbation.

Letterer-Siwe disease

- *Age*—it is fulminating condition that most often occurs in infants younger than 1 year of age.
- *Onset*—initial manifestation is skin rash involving trunk, scalp and extremities. Rash may be erythematous, purpuric and ecchymotic, some time with ulceration.
- *Symptoms*—patient may have persistent low grade spiking fever with malaise and irritability.
- *Signs*—splenomegaly, hepatomegaly and lymphadenopathy are common. Nodular and diffuse involvement of visceral organs particularly in lung and GI tract. Soft tissue and bony granulomatous reaction, hemorrhage, anemia, failure to thrive

Eosinophilic granuloma

- *Age and sex*—it occurs primarily in older children and young adults and proportion of male to female is 2:1.
- *Site*—skull and mandible are common site but femur, humerus, ribs may be affected.
- *Symptoms*—there may be local pain which may be dull and steady, swelling and tenderness may also be there. General malaise and fever may accompany the eosinophilic granuloma of bone.
- *Sign*—lesion is destructive and well demarcated, roughly round or oval in shape. The area destroyed is replaced by soft tissue. Tissue of early lesion is soft and brown and since there is no necrosis, it is not friable. Later, it become fibrous and grayish.

Oral Manifestations

Hand-Schüller-Christian disease

- *Symptoms*—sore mouth with or without ulcerative lesion, halitosis, and unpleasant taste, loose and sore teeth. There is also history of precocious exfoliation and failure of healing of tooth socket following extraction.
- *Periodontal disease*—loss of supporting alveolar bone mimics advanced periodontal disease.
- *Gingivitis*—inflammation of gingiva is also present.

Letterer-Siwe disease

- *Gingival hyperplasia*—enlargement of the gingival tissue occurs.
- *Ulcer*—there may be presence of ulcer on the oral mucosa.

- *Teeth*—diffuse destruction of bone of maxilla and mandible which may result in loosening and premature loss of teeth.

Eosinophilic granuloma

- *Site*—it may occur in jaws and overlying soft tissues.
- *Juvenile periodontitis*—there is loss of superficial alveolar bone often mimicking juvenile periodontitis.
- *Symptoms*—gingivitis and bleeding gingiva, pain or ulceration is present.
- *Signs*—loosening and sloughing of teeth often occurs after destruction of alveolar bone. Sockets fail to heal normally.

Radiological Features

Hand-Schüller-Christian disease

- *Site*—bone lesion usually seen in membrane bone but can occur in long bones and the mandible.
- *Skull lesion*—the skull lesion may be small or large, single or multiple (**Fig. 39-1**) and they may occur anywhere but anterior portion of the vault and the floor of anterior and middle cranial fossa are commonest site.

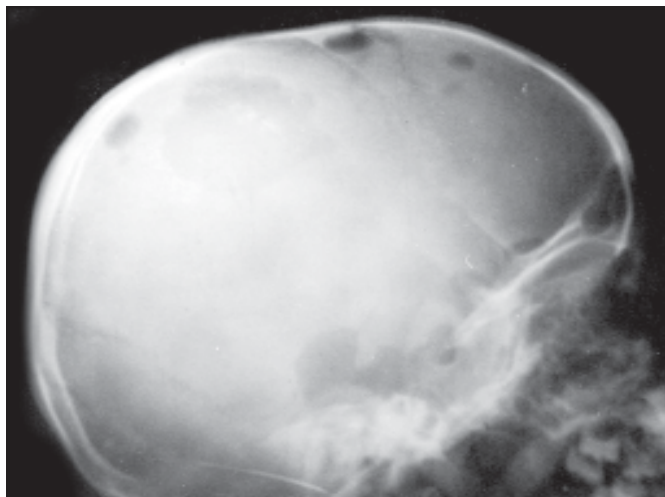


Fig. 39-1: Multiple skull lesions seen in patient with Langerhans histiocytosis.

- *Radiodensity*—the lesion is entirely destructive, so there is radiolucency which is usually sharply defined but with no cortical margin.
- *Margins*—it is well defined at periphery with punched out appearance. Margins are smooth or somewhat irregular.
- *Scooped out shape*—this particular appearance occurs because the bone destruction starts below the crest of the alveolar process (**Fig. 39-2**). Usually portions of the superior aspect of crest of the alveolar bone are maintained at the mesial and distal margins of the areas of destruction and produce scooped out appearance.

- *Floating teeth appearance*—destruction of periodontal bone support of one or more teeth especially in the posterior areas while producing virtually no resorption of tooth roots. The result is often a distinctive radiographic appearance of ‘teeth standing in space (floating teeth appearance)’, in the region superior to the mandibular canal.
- *Pathological fracture*—in some cases, there may be complete loss of continuity of the mandible, so that there is pathologic fracture.
- *Effect on surrounding structure*—displacement of erupted teeth and follicle was the most common radiographic finding. In some cases, root resorption can be seen.

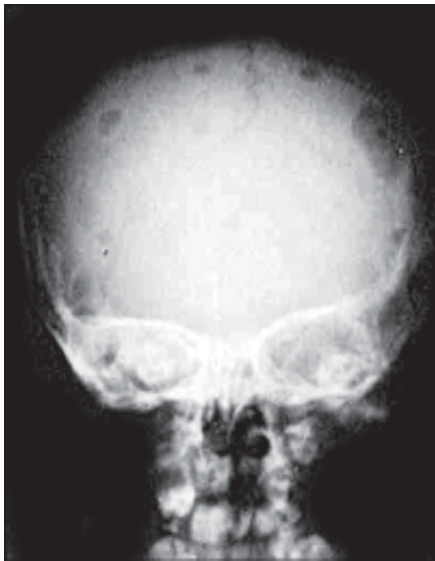


Fig. 39-2: Scooped out lesion seen in skull of the patient.

Eosinophilic granuloma

- *Site*—mandible is more commonly affected than maxilla, posteriorly more common than anteriorly.
- *Shape*—eosinophilic granuloma may be solitary or multiple and the lesions are circular or elliptical in shape.
- *Margin*—it is moderately well defined at its radiographic periphery.
- *Punched out appearance*—the lesion in the jaw have fairly discrete borders, which are rarely hyperostotic. Thus they have punched out appearance.
- *Floating teeth appearance*—jaw lesions of eosinophilic granuloma are usually seen as areas of pure osteolytic activity in close vicinity to the alveolar processes.
- *Periosteal new bone*—periosteal new bone formation is observed in some cases.
- *Bone sclerosis*—sclerosis is a common observation in inflammatory lesions of the jaws and the fact that it appears frequently in the alveolar bone lesions by communication of the lesions with the oral cavity that results in superimposed infection.

Diagnosis

- *Clinical diagnosis*—punched out destruction of skull, diabetes insipidus, ulcerative lesion, skin lesion with exophthalmos will give clue to diagnosis.
- *Laboratory diagnosis*—biopsy shows Langerhans cells which contain rod shaped cytoplasmic structure known as Birbeck granules. Plasma cells, lymphocytes, and multinucleated giant cells are also seen. Patient also suffers from anemia and less frequently leukopenia and thrombocytopenia. Serum cholesterol level is normal but tissue cholesterol level is raised.

Differential Diagnosis

- *Oral carcinoma*—found in older age.
- *Primordial cyst or dentigerous cyst*—they are more or less rounded while xanthoma is not likely to be round.
- *Benign giant cell tumor*—they usually contain some bone within the cavity while xanthoma does not. They also cause expansion of the bone. While in case of xanthoma there is no expansion of the bone.
- *Traumatic bone cyst*—they have got well defined border as compared to histiocytosis.
- *Fibrous dysplasia*—they have got clearly marked and corticated borders.
- *Apical infection*—in this case, pulp is dead.
- *Cherubism*—limited to jaw and it is bilateral.
- *Metastatic carcinoma*—incidence of histiocytosis is higher in children. A history of primary tumor helps to distinguish multiple punched out bony lesions of histiocytosis X and metastatic carcinoma.
- *Leukemia*—jaw bone lesions are most common at the crest of the alveolar ridge. But in contrast, those of leukemia characteristically originate in the deeper medullary portion.
- *Multiple myeloma*—it has to be differentiated by laboratory tests. The ratio of albumin to globulin is reserved and Bence Jones proteins are present in the urine in multiple myeloma.

Management

- *Curettage*—it is usually treated by curettage or excision of lesion.
- *Radiation*—the lesions which are inaccessible are treated by irradiation.
- *Corticosteroids*—intralesional injection of corticosteroids may be effective in local lesion.
- *Chemotherapy*—patients with acute disseminated disease, may be given drug treatment like prednisolone, vinblastine, etoposide, and cyclophosphamide.

Gaucher's Disease

It may be familial and is thought to be due to faulty metabolism of the lipid, kersin. Lack of glucocerebrosidase results in accumulation of glucosylceramide within lysosomal cells of macrophages and monocytes lineage. The reticular cells and histiocytes are increased in number and many of them become infiltrated with kersin. They become accumulate in bone marrow of this patient. It is autosomal recessive.

Types

- *Type I (chronic non-neuropathic)*—there is no cerebral involvement.
- *Type II (acute neuropathic)*—it is characterized by hepatosplenomegaly and central nervous system disorders.
- *Type III (subacute neuropathic)*—it resembles type II but later in onset and has more protracted clinical course.

Clinical Features

- *Age*—it is mostly seen in adults and progress is slow.
- *Signs*—there is enlargement of the lymph nodes, spleen and to a lesser extent the liver.
- *Symptoms*—there may be bone pain due to changes in bone marrow. There is often bleeding from the nose or from the gums. Teeth extraction from the affected area may result in bleeding complications.
- *Signs*—there is hepatosplenomegaly and CNS involvement. Patient also suffers from anemia and thrombocytopenia.
- *Pingueculae*—the skin is sometimes pigmented and the conjunctival fibrous tissue may be thickened and of brownish discoloration, a condition known as 'pingueculae'.
- *Prognosis*—the prognosis of malignant infantile form is very poor; the disease results in death usually within the first year.

Radiographic Features

- *Rarefaction*—bone changes occur due to destructive infiltration of the cerebrosidic reticulosis of the bone marrow. As a result of this and proliferation of cells so called Gaucher cells, the bone undergoes rarefaction.
- *Porosity*—there may be generalized porosity of the mandible and maxilla, with loss of trabecular structure. Sometimes, there is porosity in the mental region and thinned out mandibular cortex and areas of osteolysis present in maxillary premolar area.
- *Worm eaten appearance*—pseudocystic radiolucent areas in the molars and premolars with 'worm eaten appearance' can be seen.

Diagnosis

- *Clinical diagnosis*—pingueculae on skin, features of anemia and thrombocytopenia with hepatosplenomegaly may give clue to the diagnosis.
- *Radiological diagnosis*—worm eaten appearance with loss of lamina dura.
- *Laboratory diagnosis*—biopsy of spleen and liver show typical Gaucher cells which are round pale cells, containing a small eccentric nucleus and wrinkled or crumpled silk cytoplasm.

Management

- *Glucocerebrosidase*—enzyme replacement therapy with macrophages targeted glucocerebrosidase is used.

Niemann-Pick Disease

It is also called as '*sphingomyelin lipidosis*'. It is characterized by an abnormal storage of phospholipids due to lack of sphingomyelinase.

Clinical Features

- *Site*—it involves lungs, liver and the nervous system.
- *Signs*—there is hepatosplenomegaly, retarded physical and mental growth and severe neurological disturbance.

Diagnosis

- *Clinical diagnosis*—not so specific.
- *Laboratory diagnosis*—biopsy shows presence of foam cells, usually an enlarged reticuloendothelial cell whose cytoplasm contains numerous droplets-like inclusion of lipid.

Management

- *Symptomatic*—it is only symptomatic to control the infection. Death usually occurs within 3 years of age.
- *Others*—enzyme replacement therapy and organ transplantation are also carried out.
- *Genetic counseling*—it should be provided for the affected family.

Tay-Sachs Disease

It is caused by lack of hexosaminidase A, which results in the accumulation of ganglioside within lysosomes of neurons. This condition is heterogeneous. There is progressive neuronal degeneration developing after birth. Death occurs at 3 to 5 years of age.

Disturbances in Carbohydrate Metabolism

Classification

- *Monosaccharide*—these are those carbohydrates which cannot hydrolyze further to yield smaller carbohydrates. For example: glucose and fructose.
- *Oligosaccharides*—these are condensation product of two to three monosaccharides. For example: sucrose, maltose, lactose and raffinose.
- *Polysaccharides*—these are high molecular weight polymers of monosaccharide. For example: cellulose, starch, glycogen and dextrin.

Function

- *Energy source*—carbohydrates are the best source of energy for human body.
- *Fuel for fetus*—glucose is a major fuel of mammalian tissue and a universal fuel of fetus.
- *Cell membrane*—carbohydrates occur in cell membrane.

Hurler's Syndrome

It is disturbance of mucopolysaccharide metabolism, which is characterized by elevated mucopolysaccharide excretion ratio and an excessive intracellular accumulation of chondroitin sulphate B and heparin sulphate in those tissue and organ where they are normally found. It is inherited as an autosomal recessive trait.

Clinical Features

- *Age*—it usually becomes apparent within first two years of life, progresses during early childhood and adolescence and terminates in death before puberty.
- *Signs*—head is large, prominent forehead, broad saddle nose and wide nostrils, hypertelorism and puffy eyelids with coarse bushy eyebrows, thick lip, large tongue, open mouth and nasal congestion and noisy breathing (**Fig. 39-3**).
- *Abdomen*—progressive corneal clouding, hepato-splenomegaly resulting in protuberance of abdomen.
- *Claw hand*—a short neck and spinal abnormalities are typical, while flexion contractures result in the 'claw hand' (**Fig. 39-4**).

Oral Manifestations

- *Mandible*—shortening and broadening of mandible with prominent gonions and wide intergonial distance.
- *Teeth*—there is typical spacing of teeth. Teeth are small and misshapen.
- *Gingiva*—there may be gingival hyperplasia.



Fig. 39-3: Hurler syndrome showing large head and prominent forehead (Courtesy Dr Shetty).



Fig. 39-4: Claw hand seen in Hurler syndrome (Courtesy Dr Shetty).

Radiological Features

- *Bone destruction*—localized areas of bone destruction in the jaws may be found which appear to represent hyperplastic dental follicles with large pool of metachromate material probably mucopolysaccharide.
- *Dentigerous cyst type appearance*—radiolucent area resembles a dentigerous cyst.

Diagnosis

- *Clinical features*—open mouth appearance (**Fig. 39-5**), claw hand and puffy eyelid will diagnose this condition.
- *Radiological diagnosis*—destructive lesion of bone is seen radiologically.



Fig. 39-5: Open mouth appearance seen in Hurler syndrome (Courtesy Dr Shetty).

- **Laboratory diagnosis**—there is excessive accumulation of intracellular mucopolysaccharide in many tissues and organs throughout the body including liver, spleen, reticuloendothelial system, nervous system, cartilage, bone and heart. Abnormal deposits are found in many sites with involved fibroblasts assuming the appearance of clear (gargoyle) cells. There is elevated level of mucopolysaccharide in the urine.

Management

Death usually occurs before the age of ten due to pneumonia and cardiac failure.

Lipoid Proteinosis

It is described in Chapter 11: Keratotic and Nonkeratotic Lesions.

Hereditary Fructose Intolerance

It is transmitted as autosomal recessive trait. It results from a deficiency in fructose 1-phosphate aldolase. It is manifested by hypoglycemia and vomiting after ingestion of fructose containing foods. Affected individuals rapidly acquire an intense aversion to all sweets and fruits. There are fewer incidences of caries in these individuals.

Hypoglycemia

It is a subnormal blood glucose level. It is caused by delayed meal, inadequate total caloric intake, unusual physical

exertion, insulin measurement error, insulin overdose, the “brittle” diabetic, and oral hypoglycemic agents.

Clinical Features

- **Symptoms**—sudden onset of hunger, sweating, shakiness, tremor, anxiety, restlessness, faintness, weakness, nausea, palpitations. Progression to mental confusion, bizarre behavior, personality change, reduced level of consciousness, loss of consciousness, seizures.
- **Signs**—pulse rapid, blood pressure may be elevated and moderate hypothermia may be present due to sweating, hyperventilation and vasodilatation.
- **Other features**—bizarre or aggressive behavior may be present, staggering gait, may appear drunk, cold and clammy skin.

Diagnosis

- **Clinical diagnosis**—hunger onset, anxiety and mental confusion will give clue to the *diagnosis*.
- **Laboratory diagnosis**—blood glucose reduced.

Management

- **Consult physician**—consult physician, ensure that the airway is patent and protected and that ventilation is adequate in the patient with a reduced level of consciousness.
- **Determination of serum glucose**—determine the serum glucose with glucometer and give the conscious patient a glass of orange juice or some other form of rapidly absorbed sugar.
- **Glucose administration**—when patient is unable to take glucose orally, then give: adult—Glucagons 1 mg IM/SC, if no response may repeat in 5-20 minutes, child—Glucagons 0.025 mg/kg IM/SC, if no response may repeat in 5-20 minutes.
- **Glucagon**—glucagon is well absorbed and a response should be seen within 5 minutes. It is less sclerosing to veins than Dextrose 50% in water, start an IV with normal saline to keep vein open.

Disturbances in Mineral Metabolism

Acrodermatitis Enteropathica

It is also called as ‘zinc deficiency’. Rare disease of infancy and childhood, transmitted as autosomal recessive character. Since supplements appear to be curative even pathogenesis of the characteristic lesions involves a number of etiologic factors.

Clinical Features

- **Age**—it is usually occur in childhood after weaning.
- **Symptoms**—the primary signs of the disorder are skin lesion, hair loss, nail changes and diarrhea.
- **Signs**—erythematous, pustular, moist erosions of the orofacial areas occur as an early manifestation. In fully developed conditions, the buttocks, elbows, fingers and toes are affected by vesiculobullous rash similar to that affecting the orofacial region. Retarded body growth and mental changes also occur with some frequency.

Oral Manifestations

- **Site**—the buccal mucosa, palate, gingiva and tonsils.
- **Candidiasis**—large number of children suffer from candidiasis.
- The perioral area usually being affected by weeping erosions, angular fissuring and spreading dermatitis.
- **Papilloma**—there may be numerous small whitish papillomas on the buccal mucosa and borders of the tongue.
- The oral changes are sometimes described as 'stomatitis', 'glossitis' and 'stomatitis producing thrush like picture'.
- **Signs**—buccal mucosa is present with red and white spots, erosions, ulcers and desquamation.
- **Tongue lesion**—lesion on the tongue is sometime papillated and halitosis is often severe.

Diagnosis

- **Clinical diagnosis**—stomatitis, skin lesion, and hair loss will give clue to the diagnosis.
- **Laboratory diagnosis**—serum level of zinc is decreased.

Management

- **Zinc sulfate**—220 mg of zinc sulfate tds daily produces remarkable improvement.

Phosphorus

Phosphates form an intermediate stage in the metabolism of fats and carbohydrates by their function in phosphorylation. They are used in building the more permanent organic phosphates including some catalysts essential to the structure and function of cells. They are utilized in the formation of phosphoproteins, such as milk casein and in the formation of the nerve polypeptides. They provide the energy rich bonds such as adenosine triphosphate, which is important in muscle contraction and they form part of such coenzymes as pyridoxal phosphate, which is necessary in decarboxylation and transmission of certain amino acids such as tyrosine, tryptophan and arginate.

The suggested dietary intake of phosphorus ranges from 240 mg for infants to 800 mg for adults. It is increased in lactating and pregnant women by 50%.

Hypophosphatasia

It is a hereditary disease transmitted as a recessive autosomal characteristic. There is deficiency of serum alkaline phosphate. It resembles rickets. There is low level of serum alkaline phosphatase activity and elevated urinary excretion of phosphorylethanolamine which result in formation of defective bone matrix.

Types

- **Perinatal type**—it is diagnosed at birth and death occurs within few hours of birth. Death occurs due to respiratory failure.
- **Infantile type**—it appears at 6 months of age.
- **Juvenile type**—this appears in childhood and has wide range of clinical expression.
- **Adult type**—it is mild form and appear late in life.

Clinical Features

- **Symptoms**—there is anorexia, irritability, persistent vomiting and mild pyrexia.
- **Infantile form**—severe hypocalcemia, bone abnormalities and failure to thrive manifest the infantile form. Most of the cases are lethal. Deformities of rib may be seen in these patients.
- **Juvenile form**—hypophosphatasia of childhood is characterized by increased infection, growth retardation (**Fig. 39-6**) and rochite-like deformities including deformed extremities, costochondral junction enlargement (rochite rosary) and pulmonary gastrointestinal and renal disorders.
- **Adults form**—the adult form includes fracture with a prior history of rickets and osseous radiolucency. Stress fracture of metatarsal bones of feet may be presenting sign of this condition.
- **Homozygous involvement**—disease with homozygous involvement begins *in utero* and patient die within 1st year. Bowed limb bone and marked deficiency of skull ossification.
- **Heterozygous involvement**—in heterozygous, there is milder effect with poor growth and fracture and deformities.
- **Skull**—skull suture close early resulting in bulging suture and gray marking on internal surface of skull which occur due to increased intracranial pressure. Shape of skull is brachiocephalic.
- **Pseudohypophosphatasia**—disease resembling classic hypophosphatasia but with normal serum alkaline phosphatase level. There is osteopathy of long bones and skull. There is premature loss of deciduous teeth.

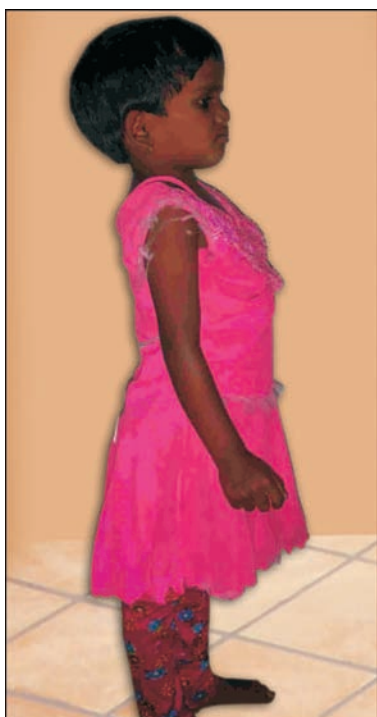


Fig. 39-6: Growth of this child is retarded due to hypophosphatasia (Courtesy Dr Manish Thadani).

Oral Manifestations

- *Premature loss of teeth and root resorption*—there is almost total lack of cementum and normally attached periodontal fiber leading to poor support and premature loss of teeth in deciduous teeth (Fig. 39-7). There is also delayed eruption of permanent teeth. The roots either fails to develop fully or undergo early resorption of the apices.
- *Hypoplasia*—teeth may be hypoplastic.
- *Pulp chamber and root canal*—the pulp chamber and root canal are sometimes larger than normal.



Fig. 39-7: Premature loss of primary teeth in child plus enamel hypoplasia is seen (Courtesy Dr Manish Thadani).

- *Alveolar bone*—the alveolar bone which support the teeth fail to develop normally which result in premature loss of primary teeth.
- *Gingiva*—there is inflammation of the gingiva.

Radiographic Features

- *Irregular ossification*—the metaphyses of long bones have been described as showing a spotty, streaky or irregular ossification.
- *Long bone*—elongated very fine processes of uncalcified tissue extend longitudinally into the poorly formed osseous tissue at the ends of long bones. Periosteal new bone formation may be present along the shaft of the long bones.
- *Skull*—skull show poor calcification.
- *Beaten copper appearances*—multiple radiolucent areas on skull called *gyral* or *convolutions* marking which described as beaten copper appearance which results due to increased intracranial pressure.
- *Jaw bone*—generalized reduction in bone density.
- *Lamina dura*—cortical and lamina dura thinning and alveolar bone deficiency.
- *Teeth*—reduced enamel thickness, enlarged pulp chamber and root canal and presence of large pulp chamber as well as alveolar bone loss.

Diagnosis

- *Clinical diagnosis*—premature loss of teeth is typical feature of this disease.
- *Radiological diagnosis*—beaten copper appearance seen and there is also thinning of lamina dura.
- *Laboratory diagnosis*—reduce level of bone, liver and kidney isozyme of alkaline phosphatase. There is also increased level of blood and urinary phosphoethanolamine. Biopsy shows abundant poorly mineralized osteoids.

Management

- *Symptomatic*—treatment of hypophosphatasia usually is symptomatic as infusion of alkaline phosphatase is not successful.
- *Prosthetic appliance*—these are given to replace missing tooth.
- *Genetic counseling*—this should be carried out to identify carriers of defective gene.

Calcium

- *Action*—it plays a large role in the formation of bones and teeth. It is also useful in the maintenance of skeletal structure, tooth structure, normal membrane permeability, normal heart rhythm and other neuromuscular

activities. It is one of the important constituent in coagulation of the blood.

- *Daily requirement*—daily recommended calcium intake of 360 mg for newborn and infants and 800 mg for children and adults. Adolescents and pregnant and lactating women are advised to increase their daily dietary calcium intake by 50%, i.e. 1200 mg.
- *Absorption and excretion*—only about 1/3rd of daily intake of calcium is absorbed under normal conditions. High protein diet has shown to increase calcium absorption. Calcium is excreted in both faeces and urine.
- *Deficiency*—deficiency may lead to internal hemorrhage and generalized paralysis can occur. There may be hyperirritability and tetany with characteristic carpopedal spasm and sometimes laryngospasm and convulsion. Derangement of blood coagulation and the integrity of the capillaries.

Magnesium

- *Action*—magnesium appears to participate practically in every phosphorylating mechanism. It is also needed for activity of certain enzymes such as phosphatase and carboxylase.
- *Daily requirement*—the recommended daily dietary allowance for magnesium ranges from 50 mg for infants to 400 mg for teenage males. A daily increase of 150 mg is suggested during pregnancy and lactation.
- *Deficiency*—deficiency of magnesium occurs due to gastrointestinal loss of the mineral resulting from frequent diarrhea and vomiting.
 - *Neuromuscular hyperirritability*—patients with deficiency exhibit severe neuromuscular hyperirritability including carpopedal spasm and a positive Chvostek sign, athetosis movements, marked susceptibility to auditory, visual and mechanical stimuli, a decreased serum magnesium level and normal calcium level.
 - *Personality changes*—there may be personality changes, anorexia, nausea and vomiting.
 - *Tetany*—the tetany appears when the serum magnesium level is depressed below 1.30 mEq per liter.
 - *Oral manifestation*—there may be localized degeneration with enamel hypoplasia of the teeth.
 - *Management of deficiency*—treatment is done by intramuscular injection of magnesium sulfate followed by a prompt rise in serum magnesium and a concomitant disappearance of the tetany and the convulsion.
- *Excess*—the administration of magnesium containing antacids to patients with renal insufficiency has resulted in central nervous system depression. There may be severe voluntary muscle paralysis. High magnesium intake will produce rickets in growing animals.

Sodium

- *Action*—sodium plays an important role in the maintenance of the acid base equilibrium as well as of osmotic pressure which depends on total base.
- *Daily requirement*—the minimum requirement of salt is thought to be about 0.5 gm.
- *Absorption and excretion*—the kidney is the principal organ for excretion of water and salt. When the diet is low in salts and when there is profuse sweating, there is no sodium found in the urine. The regulatory mechanism controlling the reabsorption of sodium by the renal tubules is controlled by adrenal gland.
- *Deficiency*—sodium deficiency occurs in man probably when diet very low in salt is used for long period of time. It is associated with gradual weakness, excessive fatigue, lassitude, apathy, anorexia, a sense of exhaustion, nausea, muscle cramps and peripheral vascular collapse.

Potassium

- *Daily requirement*—most of potassium is intracellular and average requirement of potassium is 2 to 4 gm daily. The requirement is greatest during periods of rapid growth.
- *Excretion*—about 90% of potassium is excreted in the urine.
- *Deficiency*—deficiency may occur due to loss of potassium through diarrhea and vomiting, malnutrition state, administration of diuretics and ion exchange resins, excessive dose of cortisone or hydrocortisone and diabetic acidosis during insulin therapy. The signs of potassium deficiency are primarily those of muscular irritability, muscular weakness, reduced or absent reflex, mental confusion, paralysis, disturbance in conductivity and contractility of heart muscle and alterations in the gastrointestinal tract.
- *Hyperkalemia*—it may result from extensive tissue breakdown, adrenal insufficiency, advanced dehydration on administration of excessive amounts of potassium. Hyperkalemia will produce mental confusion, numbness and tingling of the extremities, pallor, cold skin, weakness, disturbances in cardiac rhythm and peripheral collapse.

Miscellaneous Disorders

Malabsorption Syndrome

It is also called as 'sprue', 'idiopathic steatorrhea', 'celiac disease'. It includes conditions causing poor digestion or absorption to a variable degree of a number of nutrients, fats, proteins, carbohydrates, vitamins, minerals and water. The defective absorption may be due to defective digestive

or defective intestinal absorption. This disturbance in calcium metabolism may result in osteoporosis and other skeletal anomalies.

Clinical Features

- *Prodromal symptoms*—it usually begins with intestinal disturbances including diarrhea, constipation and flatulence.
- *Symptoms*—nervous irritability, numbness and tingling of the extremities occur; malaise and generalized weakness are also common.
- *Signs*—the skin changes include irregular brownish pigmentation particularly on the face, neck, arms and legs and drying of skin with scaly eruptions.

Oral Manifestations

- *Glossitis*—there may be severe glossitis with atrophy of filiform papilla, although the fungiform papillae persist for some time on the atrophic surface.
- *Symptoms*—painful burning sensation of the tongue and oral mucosa are common.
- *Signs*—there are small projections which are pink or red in color and the erythematous swelling and palatal lesions appear as multiple aphthous ulcers.

Diagnosis

- *Clinical diagnosis*—intestinal disturbances with glossitis, and erythematous swelling.
- *Laboratory diagnosis*—there is low blood calcium level.

Management

- *Vitamin*—administration of vitamin B₁₂ and folic acid.
- *Diet*—diet must be carefully supervised and supplemented with vitamins and minerals.

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Section **5**

Drugs used in Dentistry



Introduction

Antibiotics are chemical substances produced by microorganisms (fungi, actinomycetes, bacteria) that suppress the growth of other microorganisms and may eventually destroy them. Waksman and Woodruff in 1942 formally defined an antibiotic as a chemical substance produced by microorganisms, which at a high dilution can inhibit the growth and/or multiplication or kill another microorganism.

Antimicrobial drugs are the greatest contribution of the present century to therapeutics. The purpose of antibiotic/antimicrobial chemotherapy is to aid the host defenses in controlling and eliminating microbes that temporarily have overwhelmed the protective host mechanisms. Antimicrobial agents share pharmacokinetic determinants with other drugs, including ability to pass through membranes (absorption), diffuse through extracellular fluids (distribution), undergo biotransformation by hepatic enzymes (metabolism) and be eliminated from the body by renal or fecal routes (excretion).

In addition, each antimicrobial agent demonstrates unique pharmacokinetic properties including the acidic dissociation constant (pKa), lipid solubility, plasma protein binding, volume of distribution (degree of dispersal of the drug through the intracellular and extracellular fluids) and rate and type of hepatic metabolism or renal excretion. The volume of distribution and hepatic and renal excretion ratios constitute the major determinants of a drug's half-life (the time required for the peak concentration of a drug to fall by one half in a given body fluid).

The antimicrobial therapy, which targets a living organism with a physiology different from that of host, by the influence of the variables such as microorganism density, growth phase, ease of development of resistance and glycocalyx formation capable of interfering with antibiotic diffusion.

The determinants of the clinical outcome of antimicrobial therapy includes the nature and virulence of the offending organism, site of infection, possibility of establishing surgical drainage, ability of antibiotic to penetrate to the infected site, postantibiotic effects, adverse drug reactions and efficiency of the host defenses.

Since it is a unique host-parasite interaction, the unit dosages, dosing intervals and duration of therapy are not yet precisely established for the majority of specific infections.

The word '*chemotherapy*' can be defined as the use of chemical compounds in the treatment of infections, so as to destroy offending organisms or parasites without damaging the host tissues.

A chemotherapeutic agent may act by destroying the organism (bactericidal) or by inhibiting its growth (bacteriostatic). These drugs can hit at least 4 targets in bacteria: cell wall, cytoplasmic membrane, ribosomes and RNA molecules involved in transcription of genetic information.

Classification

Antimicrobial drugs can be classified in many ways:

According to Chemical Structure

- *Sulfonamides and related drugs*—Sulfadiazine and others, Sulfones-Dapsone (DDS), Para Aminosalicic acid (PAS)
- *Diaminopyrimidines*—Trimethoprim, Pyrimethamine
- *β -lactam antibiotics*—Penicillin, Cephalosporin
- *Tetracycline*—Oxytetracycline, Doxycycline
- *Nitrobenzene derivative*—Chloramphenicol
- *Aminoglycosides*—Streptomycin, Gentamycin, Neomycin etc
- *Macrolide*—Erythromycin, Oleandomycin
- *Polypeptide antibiotics*—Polymyxin-B, Colistin, Bacitracin, Tyrothricin

- Nitrofurans derivative—Nitrofurantoin, Furazolidone
- Nitroimidazoles—Metronidazole, Tinidazole
- Quinolones—Nalidixic acid, Cinoxacin, Rosaxacin
- Nicotinic acid derivative—Isoniazid, Pyrazinamide, Ethionamide
- Polyene antibiotics—Nystatin, Amphotericin B, Hamycin
- Imidazole derivative—Miconazole, Ketoconazole, Clotrimazole
- Others—Rifampin, Clindamycin, Spectinomycin, Vancomycin, Lincomycin, Sodium fusidate, Cycloserine, Viomycin, Ethambutal, Thiacetazone, Clofazimine, Griseofulvin.

According to Mechanism of Action

- Interference with cell wall synthesis—Penicillin, Cephalosporin, Bacitracin, Vancomycin and Cycloserine C
- Damage to cytoplasmic membranes—Polypeptides: Polymyxins, Colistin, Bacitracin
- Polyene antibiotics: Amphotericin B, Nystatin, Hamycin
- Inhibition of protein synthesis and impairment of function of the ribosomes—Aminoglycosides, Tetracycline, Chloramphenicol, Macrolide and lincomycin
- Interference with transcription/ translation of genetic information (misreading of mRNA code)—Quinolones, Metronidazole, Rifampin and Ethambutol
- Antimetabolite action—Sulfonamides, Sulfones, PAS and Trimethoprim
- Binding to viral enzymes essential for DNA synthesis (Interfere with DNA function)—Vidarabine, Acyclovir

According to Type of Organism against which Primarily Active

- Antibacterial—Penicillin, Aminoglycosides, Erythromycin etc
- Antifungal—Griseofulvin, Amphotericin B, Miconazole
- Antiviral—Idoxuridine, Acyclovir, Amantadine
- Antiprotozoal—Chloroquine, Pyrimethamine, Metronidazole, Diloxamide
- Anthelmintic—Mebendazole, Piperazine, Pyrantel, Niclosamide.

According to Spectrum of Activity

- Narrow spectrum—Penicillin G, Streptomycin, Erythromycin
- Broad spectrum—Tetracycline, Chloramphenicol

According to Type of Action

- Primary bacteriostatic—Sulfonamides, Tetracycline, Chloramphenicol, Erythromycin, Ethambutol

- Primary bactericidal—Penicillins, Cephalosporins, Aminoglycosides, Polypeptides, Rifampin, Cotrimoxazole, Vancomycin, Nalidixic acid, Isoniazid (Some primary 'static drugs' may become 'cidal' at higher concentrations, e.g. Sulfonamides, Erythromycin etc.; On the other hand some 'cidal' drugs may only be static under certain circumstances, e.g. Cotrimoxazole, Streptomycin).

According to Antibiotics Obtained From

- Fungi—Penicillin, Cephalosporin, Griseofulvin
- Bacteria—Polymyxin B, Colistin, Bacitracin, Tyrothricin
- Actinomycetes—Aminoglycosides, Tetracyclines, Chloramphenicol, Macrolides, Polyenes
- Entirely by chemical method—Chloramphenicol

According to the Organisms Susceptible

- Antibiotics mainly effective against Gm +ve bacteria—For systemic infections, e.g. Penicillins, Macrolides. For topical use, e.g. Bacitracin
- Antibiotics mainly effective against Gm-ve bacteria—For systemic, e.g. Streptomycin and other Aminoglycosides
- Antibiotics effective against both Gm +ve and Gm-ve bacteria—For systemic infections: Ampicillin, Amoxicillin, Cephalosporins, etc. For topical use: Neomycin, Framycetin
- Antibiotics effective against both Gm +ve and Gm-ve bacteria: Rickettsiae and Chlamydia—Tetracyclines, Chloramphenicol
- Antibiotics effective against acid-fast bacilli (*Mycobacterium tuberculosis*)—Streptomycin, Rifampin, Kanamycin
- Antibiotic effective against protozoa—Tetracycline
- Antibiotic effective against fungi—Nystatin, Amphotericin B, Griseofulvin
- Antimalignancy antibiotics—Actinomycin D, Mitomycin.

Routes of Drug Administration

Route and time of administration are major factors in determining ultimate concentration at the site of infection.

Antimicrobial drugs can be administered:

- Topically/ Locally
- Orally
- Intramuscular injection
- Intravenous injection

With few exceptions, the absorption of antibiotics given orally is decreased in the presence of food. Some antibiotics are acid unstable and nearly complete inactivation of these drugs will occur if it is taken just before and after meals when gastric secretions are high. Therefore, in general, antibiotics should be taken no sooner than 1 hour before or

at least 2 hours after a meal. In addition, interaction with other orally administered drugs can decrease the external absorption of some antibiotics. For example, the absorption of tetracycline is decreased by milk, by antacids containing divalent and trivalent metallic cations and by medications containing iron salts.

Antimicrobial Dosing Principle

The proper dose of a drug is the amount that produces the maximum benefit, with the least attendant harm, i.e. with minimal adverse effects on the physiology of the host and the microbial ecology.

- *Vigorous dosage*—the current trend in antimicrobial therapy is to give a vigorous dosage for a short time as the clinical situation permits. Such a regimen is desirable since a major factor in the success of most antimicrobial agents is the height of the concentration in the serum and in the infected tissue by the drug. High concentrations are more critical with aminoglycoside, metronidazole and quinolones (concentration-dependent antibiotics), whereas prolonged exposure of the organism to the antimicrobial agent is more critical with β -lactams (time-dependent antibiotics). Prolonged dosing beyond what is necessary increases the antibiotic toxicity, allergy and selection of resistant microorganisms.
- *Loading dose*—use of an oral antibiotic loading dose: Without a loading dose, it takes 6-12 hours to achieve maximum therapeutic blood and tissue levels via oral administration.
- *Achieving maximum blood level*—achieve blood levels of the antibiotic at 2-8 times the minimal inhibitory concentration. Such blood levels are necessary to compensate for the tissue barriers that impede antibiotic penetration to the site of the injection.
- *Use frequent dosing intervals*—this is important with the older β lactam antibiotics such as penicillin V and the first generation cephalosporins so as to maintain relatively constant blood levels.
- *Determine duration of therapy by remission of disease*—the antimicrobial agents is terminated when the patient host defenses have gained control of the infection and the infection is reasonably certain to resolve or has resolved.
- *Improper dosing*—it may be in 3 forms:
 - Inadequate concentration of drugs
 - Inadequate duration of therapy
 - Excessive dosage (leading to toxic effects in the host).

Duration of Antibiotic Therapy

A significant misconception in antibiotic therapy is that antibiotic use requires a “complete course” of therapy.

Conceptual errors about a preordained “course” of antimicrobial therapy emanate from several faulty assumptions:

- *Resistant microorganism*—prolonged antimicrobial therapy destroys resistant microorganisms. This is a contradiction in terms, as antimicrobial agents cannot affect microorganism resistant to them and the prolonged use of antimicrobial agents only serves to select for these resistant species.
- *Rebound infection*—prolonged antibiotic therapy is necessary for “rebound” infections that recur since the organisms are suppressed but not eliminated. Acute orofacial infections do not rebound particularly if the source of the infection is probably eradicated.
- *Variability in infectious process*—antibiotic dosages and duration of therapy can be extrapolated from one infection to another. This is not possible to give the variability in infectious processes.
- *Remission and relapse of infection*—according to Norsby S. R (1991) the ideal duration of antibiotic therapy is the shortest that will prevent both clinical and microbiological relapse. According to Leitman P. S (1990), the only practical guide for determining the effectiveness of antimicrobial treatment and hence the duration of therapy is clinical improvement of the patient as judged by remission of the infection. Acute orofacial infections have a rapid onset and relatively short duration of 2-7 days or less, particularly if the offending cause is treated or eliminated. If clinical experience and the nature of the infection dictate that it is anticipated course, if it will be 3 days, then 3 days of antibiotic therapy is enough; if 5 days, then 5 days of therapy, etc. When clinical evidence indicates that the infection is reasonably certain to resolve or is resolved, the antibiotic therapy should be terminated.
- *Rule of thumb*—according to the rule of thumb, antibiotic coverage should last for at least 48 hours after complete remission of clinical symptoms. Treatment of the usual oral infections of bacterial origin requires an average of 5-7 days of antibiotic therapy.

Effectiveness of Antibiotics

Age, Type and Extent of Infection

The time duration of the presence of an infection plays a significant role in determining the effectiveness of antibiotic therapy. Older, well established infections grow slowly and thus both bactericidal and bacteriostatic antibiotics are less effective than they would be in new infections.

The larger the size or extent of infection, the less effective the antibiotic therapy will be, because the increased number of bacteria generally requires a higher concentration of antibiotic be present at the site of infection.

Antibiotics by themselves cannot be relied on to eradicate some infections. Appreciable quantities of pus, necrotic tissue, a foreign body and abscessed areas that are relatively avascular require prompt surgical management in addition to adequate doses of antimicrobial agents.

Resistance

Some species of bacteria are protected against the action of certain antibiotics by virtue of the fact that the drug will not reach the target site (e.g. cell wall may be impermeable). Also, bacteria that would normally be expected to be susceptible to a particular antibiotic can develop resistance. Resistance to antimicrobials can be produced by a number of mechanisms.

- *Decreased permeability*—By the decrease in the permeability of the bacterial cell membrane to the drug (e.g. resistance to ampicillin).
- *Inactivation of drug*—by inactivation of the drug by enzymes of bacteria (e.g. Betalactamase inactivates some penicillin).
- *Alteration of drug receptor*—by alteration of the drug receptor site in bacteria (e.g. in erythromycin, there is a change in the ribosomal binding site).
- *Alternative metabolic pathway*—by the development of an alternative metabolic pathway in bacteria which are unaffected by the drug (e.g. in sulfonamide resistance).
- *Increases elimination of drug*—by increased elimination of drugs from the bacterial cell (e.g. Fluoroquinolones may be actively expelled from the cell).
- *Mutation*—resistance can develop from one bacterial generation to the next by mutation (vertical transfer). Mutation is not a result of exposure to antibiotics but there are spontaneous mutations occurring continually in the bacterial population and some of them will coincidentally confer resistance.

Patient Compliance

This is a very important factor since patients may miss doses when clinical symptoms begin to subside and a patient may choose to discontinue therapy as soon as overt symptoms are absent. Premature termination of drug therapy will favor selection of microorganisms that are more resistant to the drug. Particularly with bacteriostatic agents, patient compliance is a requisite for a successful clinical outcome.

Concentration of Antibiotics at the Site of Infection

To be effective, an antibiotic must reach the site of infection in amounts above the MIC for infecting microorganism. (MIC—Lowest concentration needed to inhibit visible growth of an organism on media after 18-24 hours of incubation).

Metabolism and Excretion of Antibiotics

Alteration of a drug within a living organism is known as biotransformation. The time during which an antibiotic is present in effective concentration at the site of infection depends on the rate of inactivation of the drug through metabolism or excretion or both. Based on this, the dose is repeated at specific intervals so that antibacterial blood concentration is maintained. After absorption, drugs can undergo 3 possible fates:

- Metabolic degradation by enzymes (by oxidation, reduction or hydrolysis)
- Spontaneous change into other substances
- Excretion as the same or unchanged.

Penicillins, cephalosporin, aminoglycoside and polymyxins are excreted mainly through kidney. Other antibiotics undergo varying degree of biotransformation in the liver.

Indications for Antibiotics

Therapeutic

- In patients where the host response is decreased by diseases like diabetes mellitus, immunoglobulin deficiency, malnutrition and alcoholism.
- Acute severe rapidly spreading infection.
- Pericoronitis, osteomyelitis, fracture, soft tissue wound and odontogenic infection.

Prophylactic

- More than 1/3rd are used for this purpose.
- For postoperative wound infection.
- In prevention of endocarditis in high risk patients undergoing any dental procedures that likely to cause gingival bleeding.

Inhibition of Protein Synthesis

Tetracyclines

The first member of this group called *chlortetracycline* and the second member *oxytetracycline* are derived from streptomyces species. Later in 1953, tetracycline has been prepared by catalytic hydrogenation of chlortetracycline. (Chemically, the Tetracyclines are naphthalene derivatives. The naphthalene nucleus is made up by fusion of 4 partially unsaturated cyclohexane radicals and hence the name tetracycline). Broad-spectrum bacteriostatic antibiotic was isolated from soil, i.e. Actinomycetes in 1948.

- *Action*—it inhibits protein synthesis by binding to 30S ribosomes in susceptible organism. It is effective against

a variety of gram -ve and gram +ve bacteria, rickettsial, spirochetes, protozoa, and mycoplasma. It is mainly given orally, secreted in saliva, concentrated in liver and excreted in bile. They cross placental barrier.

- *Use in dentistry*—it is used in number of dental-related conditions such as prevention of subacute bacterial endocarditis, as an adjunct to conventional periodontal therapy, for the treatment of ANUG and dental abscesses.
- *Dosage*—Tetracycline (orally 250/500 mg qid), Minocycline (orally 200 mg bid/od) and Doxycycline (orally 100 mg bid for the 1st day, with a subsequent daily dose of 100 mg OD).
- *Adverse reaction*—
 - *GIT disturbances*—it can anorexia, vomiting, heart burn, diarrhea, epigastric pain, gastroenteritis.
 - *Hepatic and renal toxicity*—there is also hepatic toxicity and renal toxicity can occur.
 - *Tooth discoloration*—hypoplasia and brown discoloration of teeth can occur; hence it is not used in 3rd trimester. All Tetracyclines are deposited in calcifying areas of the bones and teeth and may cause a yellow discoloration. Tetracyclines are not recommended for children under the age of 8 years old and pregnant women since they can permanently discolor developing teeth and alter bone growth.
 - *Increases blood urea level*—tetracycline reduces protein synthesis and has an overall catabolic effect. They induce negative nitrogen balance and can increase blood urea level.
 - *Superinfection*—Tetracyclines are the most common antibiotics responsible for superinfection.
 - *Other*—Minocycline causes dizziness, vertigo and tinnitus with high dosage. Other adverse effects, though rare, include nephrotoxicity, mild leukopenia, neurotoxicity and possibly decreased phagocytic activity.
- *Contraindication*—All tetracyclines are contraindicated during pregnancy since the drugs cross the placenta and form a stable calcium complex in bone forming tissue which can result in decrease in the fibula growth rate and retardation of skeletal development. The drug should not be given to nursing mothers since they are excreted in milk.
- *Absorption, fate and excretion*—they are not destroyed by the gastric acid or the intestinal flora. The tetracycline form insoluble complexes by chelation with calcium, magnesium, aluminium and hence substances like milk that contain calcium and antacids reduce their absorption. Administration of iron also interferes with absorption. Food also interferes with the bioavailability. The tetracycline is mainly absorbed from the duodenum and upper small intestine. Hence with larger doses, more

drugs escapes through absorption causing GIT disturbances. Tetracyclines are metabolized in the liver and metabolites are excreted mainly in the urine by glomerular filtration.

- *Properties*—Tetracyclines also have non-bactericidal properties that include anti-inflammatory, immunosuppressive properties, suppression of antibody production in lymphocytes, reduction in phagocytic function of PMNs, reduction of leukocyte and neutrophils chemotaxis, as a sclerosing agent, as an inhibitor of lipase and collagenase activity, as an enhancer of gingival fibroblast cell attachment and even at tumor activity.
- *Drug interaction*
 - *Oral contraceptive*—concurrent use of any tetracycline may render oral contraceptives less effective and may induce bleeding.
 - *Antacid*—the absorption of tetracycline from the stomach is impaired by divalent cations due to binding by the tetracycline and can decrease bioavailability. Therefore, the following should be avoided during tetracycline therapy: antacids containing aluminum, calcium or magnesium, dairy products such as milk, cheese, etc. that contain calcium and iron preparations.
 - *Penicillin*—since any bacteriostatic antibiotic may interfere with bactericidal action of penicillin, tetracycline class drugs should not be used in conjunction with any penicillin drugs.
 - *Anticoagulant therapy*—because tetracycline has proven to depress plasma prothrombin activity, patients on anticoagulant therapy (Coumarin, heparin, protamine) may require decreasing the dosage of anticoagulant drug.
 - *Methoxyflurane*—the concurrent use of tetracycline and anesthetic, methoxyflurane is contraindicated since the combination has been reported to result in fatal renal toxicity.
 - *Other drugs*—carbamazepine, Phenytoin, barbiturates, chronic ethanol ingestion may decrease the half life of tetracycline.

Macrolide

The Macrolides are a group of antibiotics that have in common macrocyclic lactone rings linked with amino-sugars. Apart from erythromycin, clindamycin, spiramycin, roxithromycin, clarithromycin and azithromycin are used clinically. Azithromycin is from a novel class of antibiotics called azalides which have been derived from Macrolides.

Erythromycin

It is isolated from streptomyces erythreus in 1952.

- **Action**—it is bacteriostatic and interferes with protein synthesis by inhibiting enzyme transferase at the 50S subunit of ribosome. It is effective against gram +ve and gram -ve bacteria. Depending upon its concentration, the antibiotic can be bacteriostatic or bactericidal. The drug is effective against penicillin resistant staphylococci.
- **Use in dentistry**—it is used as prophylactic agent against endocarditis before dental procedures and in patients allergic to penicillin. It is also used in tetanus, diphtheria and syphilis.
- **Excretion**—it is excreted through urine, faeces and bile.
- **Adverse reactions**—there may be stomatitis, abdominal pain, nausea, vomiting, jaundice, diarrhea, urticaria, and anaphylaxis. Severe adverse effects are uncommon. Hepatic dysfunction is a relative contraindication for erythromycin therapy. Super infection with Gm-ve organisms and candida may develop.
- **Dose**—it is given orally in dose of 250 to 500 mg/8 hourly.
- **Spectrum**—erythromycin has a wide range of activity against both Gm +ve facultative and anaerobic bacteria like Streptococcus, Staphylococcus, Eubacterium, Propionibacterium, bifido-bacterium, Lactobacillus and Peptostreptococcus species. However, most Gm-ve bacteria are resistant to erythromycin except Hemophilus and Neisseria species due to its inability to penetrate the lipopolysaccharide cell wall complex.
- **Absorption, fate and excretion**—the drug is partially destroyed by the gastric juice and therefore, has to be administered in the form of enteric coated tablets. The ester form, erythromycin esteolate is more resistant to inactivation by the gastric acid and is absorbed better, giving higher and more prolonged blood levels than the base. The drug can pass into fetal circulation.
- **Drug interaction**—Reduction in plasma clearance with increased plasma levels and increased toxicity when astemizole, terfenadine, theophyllin, carbamazepine, cyclosporine digoxin and warfarin are coadministered. It interferes with the action of chloramphenicol and clindamycin. Physical/chemical incompatibility between parenteral forms of erythromycin and vitamin B complex, ascorbic acid, tetracycline, heparin and phenytoin.

Other Macrolides

Azithromycin is active against Gm-ve anaerobes and provides higher drug concentrations in the tissues than in blood or serum. It has been found to be highly effective against all serotypes of Actinobacillus actinomycetemcomitans (Aa) and against Porphyromonas gingivalis. Effectiveness of erythromycin against viridans streptococci is poor. Other Macrolides, such as azithromycin and spiramycin have been used in treating dentoalveolar infections.

Azithromycin is more effective than spiramycin and might serve as an alternative antibiotic against odontogenic infections and periodontal abscess.

Aminoglycoside

- **Spectrum**—aminoglycosides are compounds that are composed of at least one sugar attached to one or more amino groups. Their main antibacterial spectrum is against Gm-ve bacilli. Some of the aminoglycoside are active against staphylococci, but most Gm +ve and most anaerobic bacteria are resistant. All are potentially ototoxic and may produce deafness due to their effect on the 8th cranial nerve.
- **Drug includes**—this group includes Streptomycin, Gentamycin, Tobramycin, Sisomycin, Kanamycin, Amikacin, Paromomycin, Framycetin and Neomycin.
- **Mechanism of action**—evidence indicates that the bactericidal effect of these drugs may be due to their ability to cause detachment of the ribosomal complex from the mRNA, which would result in the inability of the cell to produce essential proteins. The aminoglycosides are inactive under anaerobic conditions because intracellular transport is severely impaired in the absence of oxygen. Therefore, all anaerobic bacteria are markedly resistant even though they contain ribosomes that are sensitive to these antibiotics.
- **Dosage**—the dosage of these antibiotics required for clinical effectiveness is only slightly below the levels that elicit toxicity and potential adverse reactions can be serious and irreversible. Ototoxicity and nephrotoxicity have been frequently associated with use of the aminoglycoside with an incidence rate of 2-10%. Therefore, use of an aminoglycoside is somewhat limited. Due to the synergistic effect with other antibiotics, aminoglycoside are usually given in lower doses in combination with other drugs.
 - **Gentamycin**—IM/ slow IV dose (2-5 mg/kg/day in 3 divided doses) (it can produce bactericidal effect)
 - **Tobramycin**—IM/IV 3-5 mg/kg/day in 3-4 equal doses (bactericidal)
 - **Streptomycin**—0.5 to 1 gm 12 hourly for 7 days in acute infection.
- **Use**—Penicillin and Gentamycin are commonly used together for the treatment of bacterial endocarditis. Most of the aminoglycosides are only available for parenteral administration; however, some are available as creams or ointments for topical application.

Inhibitors of Cell Wall Synthesis

Penicillin

Penicillin, the most important antibiotic, was first extracted (1928) from the mould penicillium notatum. Subsequently,

a mutant of a related mould, *P. chrysogenum*, was found to give the highest yield of penicillin and is now employed for commercial production of this antibiotic. Penicillins belong to a group of antibiotics called as beta-lactams. The other members include cephalosporin, cephamycin, monobactams and carenems.

Penicillins are a family of antibiotics, with bactericidal activity against broad spectrum bacteria. The basic structure of the penicillin consists of thiazolidine ring fused with a beta lactam ring which is essential for antibacterial activity. The 2 rings constitute the fundamental nucleus of all the penicillins, namely 6-amino-penicillanic acid (6APA). Substitutions and modifications on the acyl side chain have resulted in a number of semisynthetic penicillins with enhanced antimicrobial properties such stability to gastric acids, resistance to hydrolytic enzymes and increased absorption from the stomach.

The side chain also determines the stability of the penicillin against degradation by gastric acid and by enzyme penicillinase (beta lactamase) produced by certain microorganisms. (The beta lactamase producing organisms are *Staphylococcus aureus*, *Pseudomonas*, *Enterobacter*, etc). The first identified penicillin is benzyl penicillin-G. If phenoxyacetic acid was added to the culture, phenoxy-methyl penicillin (penicillin V) was produced which is semi-synthetic one.

Classification

- *Natural penicillins*—Penicillin G (Benzyl penicillin), Procaine penicillin G and Benzathine penicillin.
- *Semi-synthetic penicillin*—
 - *Acid resistant penicillins*—phoxymethyl penicillin (Penicillin V) and Phenoxyethyl penicillin (Phenethicillin)
 - *Penicillinase-resistant penicillins*—Acid labile (Methicillin, Nafcillin, Cloxacillin, Dicloxacillin) and Acid resistant (Flucloxacillin)
- *Penicillins effective against Gm +ve and Gm-ve organisms*—Ampicillin, Amoxicillin.
- *Extended spectrum penicillins*—Carboxy penicillin (carbenicillin, Ticarcillin), Ureidopenicillins—Piperacillin, Amidinopenicillin (Mecllinam).
- *Penicillins with beta lactamase inhibitors*—Amoxicillin—Clavulanic acid (Augmentin), Ticarcillin—Clavulanic acid (Timentin).

Cloxacillin

- *Activity*—this is Penicillinase resistant penicillin. This drug has weaker antibacterial activity than penicillin G. So, it is used in conjunction with ampicillin or amoxicillin to enhance the synergism.

- *Absorption*—food interferes with the absorption of the drug. High concentration occur in kidney and liver; approximately 90 to 95% bound to plasma proteins.
- *Dosage*—the initial dose varies from 0.5 to 1 gm qid and the maintenance dose is 250 mg qid. The drug should be administered 1 hour before or 2 hours after a meal to ensure adequate absorption. It can also be given IM and by slow IV 250-500 mg 4-6 hourly.

Ampicillin

- *Action*—it is effective against Gm +ve and Gm-ve organisms. The antibacterial activity of ampicillin is generally similar to that of Benzyl penicillin, but it is more effective than benzyl penicillin against a variety of Gm-ve bacteria. The Gm +ve cocci are less sensitive to ampicillin than to benzyl penicillin. Ampicillin is inactivated by penicillinase.
- *Absorption*—it is incompletely absorbed on oral administration. Food does not interfere with its absorption. Plasma levels are reached at peak within 2 hours and 1 hour after oral and IM administration respectively.
- *Adverse effect*—ampicillin causes skin rash is usually maculopapular and not urticarial. Diarrhea is common with oral ampicillin.
- *Dosage*—the ampicillin capsule contains ampicillin hydrate equivalent to 250 mg of the base. The usual adult dose of ampicillin is 250 to 500 mg 6 hourly (qid); doses as large as 1 gm qid may be required for more refractory Gm-ve infections. High doses for IM/ IV injection are used in the treatment of meningitis and bacterial endocarditis.

Amoxicillin

- *Action*—this is effective against Gm +ve and Gm-ve organisms. This is a semisynthetic penicillin (amino-p-hydroxy-benzyl penicillin) with a broad spectrum of antibacterial activity and having substantial advantages over ampicillin.
- *Absorption*—amoxicillin is effective on oral administration and the blood levels are twice as high as those after similar dose of ampicillin. Its absorption is not affected by food. The incidence of skin rashes and diarrhea is less than with ampicillin.
- *Dosage*—250-500 mg (tid) in adult. It can also be given IV / IM.
- *Adverse reaction*—penicillins are among the least toxic of any antibiotics. Hypersensitivity is by far the most common adverse reaction noted with any penicillin. According to Idsoe et al 0.6-10% of patients treated with penicillin develop some form of allergic reactions such

as maculopapular rash, urticaria, fever, bronchospasm, vasculitis, serum sickness, exfoliative dermatitis and anaphylaxis. Mild reactions are often limited to a rash or skin lesions located on or about the head and neck that may be accompanied by facial swelling. More severe reactions may involve the joints and result in pronounced swelling or tenderness. In highly sensitized patients, anaphylaxis may occur and can result in death. IV administration has been associated with more severe reactions. Penicillin G can become more allergic after being in solution for a while, either because it breaks down to form more allergenic substances or due to the formation of high molecular weight polymers. Therefore, only freshly prepared solutions should be used. It should always be remembered that an allergy to one penicillin exposes the patient to a greater risk of reaction if another penicillin or cephalosporin is given. Broad-spectrum penicillins may alter the natural bacterial flora of the oral cavity and GIT resulting in superinfection with resistant bacteria, colonization by opportunistic pathogens, fungal infections or pseudomembranous colitis.

- **Drug interaction**—concurrent therapy of penicillin and aminoglycosides are not advised since the former may inactivate the latter.

Newer Penicillins

- **Azlocillin**—it is an acyl ampicillin antibiotic with an extended spectrum of activity and greater *in vitro* potency than the carboxy penicillins. Azlocillin is similar to mezlocillin and piperacillin. It demonstrates antibacterial activity against a broad-spectrum of bacteria, including *Pseudomonas aeruginosa* and, in contrast to most cephalosporin, exhibits activity against enterococci.
- **Dicloxacin**—it is a narrow spectrum beta-lactam antibiotic of the penicillin class. It is used to treat infections caused by susceptible Gram-positive bacteria. Notably, it is active against beta-lactamase-producing organisms such as *Staphylococcus aureus*, which would otherwise be resistant to most penicillins. It is very similar to flucloxacillin and these two agents are considered interchangeable. Dicloxacin is commercially available as the sodium salt **dicloxacin sodium** in capsules (250 or 500 mg) and injections (powder for reconstitution, 500 and 1000 mg per vial).
- **Flucloxacillin**—it is a narrow spectrum beta-lactam antibiotic of the penicillin class. It is used to treat infections caused by susceptible Gram-positive bacteria. Nowadays, it is no longer recommended against beta-lactamase-producing organisms such as *Staphylococcus aureus*, since like in other penicillins, it is not active against such infections. It is very similar to dicloxacin and these two agents are considered interchangeable.
- **Flucloxacillin** is commercially available as the sodium salt **flucloxacillin sodium**, in capsules (250 or 500 mg), oral suspensions (125 mg/5 mL or 250 mg/5 mL), and injections (powder for reconstitution, 250, 500 and 1000 mg per vial).
- **Mecillinam**—it is an extended-spectrum penicillin antibiotic that binds specifically to penicillin binding protein 2 (PBP2), and is only considered to be active against Gram-negative bacteria. It is used primarily in the treatment of urinary tract infections, and has also been used to treat typhoid and paratyphoid fever.
- **Nafcillin**—it is a narrow spectrum beta-lactam antibiotic of the penicillin class. As a beta-lactamase-resistant penicillin, it is used to treat infections caused by Gram-positive bacteria, particularly species of *Staphylococci*, that are resistant to other penicillins.
- **Oxacillin**—it is a narrow spectrum beta-lactam antibiotic of the penicillin class. It is effective against penicillinase enzymes, such as that produced by *Staphylococcus aureus*. However, resistant strains are now emerging that are called oxacillin-resistant *Staphylococcus aureus*.
- **Piperacillin**—it is an extended spectrum beta-lactam antibiotic of the ureidopenicillin class. It is normally used together with a beta-lactamase inhibitor. The combination has activity against many Gram-positive and Gram-negative pathogens and anaerobes, including *Pseudomonas aeruginosa*. Piperacillin is not absorbed orally, and must therefore be given by intravenous or intramuscular injection; piperacillin/tazobactam is administered intravenously every 6 or 8 hours; the drug may also be given by continuous infusion, but this has not been shown to be superior.
- **Ticarcillin**—it is a carboxypenicillin. It is almost invariably sold and used in combination with clavulanate. Because it is penicillin, it also falls within the larger class of beta-lactam antibiotics. Its main clinical use is as an injectable antibiotic for the treatment of gram negative bacteria, in particular, *Pseudomonas aeruginosa*. Ticarcillin is not absorbed orally, and therefore must be given by intravenous or intramuscular injection. The usual adult dose of Ticarcillin is 3.5 gm four times a day.

Cephalosporins

In 1945, Prof. G. Brotzu isolated a fungus called as *Cephalosporium acremonium*. Cephalosporins are extracted from this fungus. Cephalosporins have 7-amino cephalosporanic acid nucleus which bears close resemblance to the 6-APA nucleus of penicillins.

- **Antibacterial activity**—cephalosporins possess a wide range of activity against Gm +ve and Gm-ve bacteria. Cephalosporins act by inhibiting bacterial cell wall synthesis and are bactericidal.

- *Absorption, fate and excretion*—cephalosporins are administered either orally or IV (IM is painful). Cephalosporins are eliminated mainly by renal excretion (As with penicillins, renal tubular blocking with probenecid increases the plasma levels significantly).
- *Adverse reaction*—it can cause allergy such as skin rashes, fever, serum sickness, anaphylactic reaction (Rare), eosinophilia, neutropenia, transient splenomegaly and increased SGOT levels. Some of the newer cephalosporins cause disulfiram like effect when combined with alcohol.
- *First generation cephalosporins*—the first cephalosporin introduced into medical practice was cephalothin. This compound is active against both penicillin-sensitive and resistant staphylococci, pneumococci, streptococci and the common Gm-ve pathogens, but they are not effective against anaerobes. Other 1st generation cephalosporins are cephacetrile and cefazolin. The 1st orally active cephalosporin for medical use was cephaloglycin. Cephalexin is more stable and has better oral absorption. It is often used as follow-up therapy and for the treatment of minor infections involving Gm+ve organisms. Cephradine has a similar Gm+ve spectrum, but is more active against Gm-ve organisms.
- *Second generation cephalosporins*—these offer a wide range against Gm-ve bacilli. Their main use would appear to be in the Gm-ve infections, especially those caused by β lactamase producing organisms. But they have less activity against most pathogens that cause infection of wounds.
- *Third generation cephalosporins*—they have poor activity against the Gm+ve cocci; but are more active against Gm-ve bacilli than the first and second generation cephalosporins. This generation includes ceftazidime, cefsulodin, ceftriaxone (once a day).
- *Fourth generation cephalosporins*—Cefepime is more resistant to some β lactamase. It is active against streptococci, and methicillin-sensitive staphylococci. Its main use is in serious Gm-ve infections including infections of the CNS into which has excellent penetration. Dose: 2 gm IV bid.
- *Fifth generation cephalosporins*—this is recently discovered cephalosporin and it is used in severe infection. Example of this generation cephalosporin is ceftobiprole.
- *Cefazolin*—parenteral administration results in higher blood level that persists longer. It has a half life of 2 hours due to slower tubular secretion. Dose—0.25 gm 8 hourly (mild cases), 1 gm 6 hourly (severe cases) IM and IV.
- *Cephalexin*—it is effective against most gram +ve cocci, bacilli, *E. coli* and *Klebsiella*. It is an orally effective first generation cephalosporin. Dose—0.25 to 1 gm 6-8 hourly and in children 25-100 mg/kg/day.
- *Cefadroxil*—it is close congener of cephalexin, has good tissue penetration and it exerts more sustained action at the site of infection. Dose—0.5 to 1 gm BD.
- *Cefaclor*—it retains significant activity by the oral route. It is more active *H. influenzae*, *E. coli*. Dose—125 to 250 mg daily.
- *Cefuroxime*—it is highly active against ampicillin resistant *H. influenzae*. It is well tolerated by the IM route and attains relatively higher CSF level. Dose—0.75 to 1.5 gm IM 8 hourly.
- *Cefixime*—it is an orally active and is highly active against enterobacteria. Dose—200 to 400 mg BD.
- *Ceftobiprole*—the adult dose of ceftobiprole is 500 mg every eight hours given intravenously. Ceftobiprole cannot be given by mouth. Ceftobiprole is not licensed to be used in children.

Interference with Cell Membrane Structure

5

The selective permeability of the cytoplasmic membrane allows the bacterial cell to control the entry and exit of substances by specific transport mechanisms. This membrane is associated with a number of vital cell functions such as DNA replication and cell wall synthesis. Any agent that adversely affects the function of this membrane is usually lethal for the cell. Unfortunately, the antibiotics that exert their effect against the cell membrane do not exhibit selective toxicity and are equally destructive to both bacterial and mammalian cells.

Two general classes of antibiotics belong to this category, 1) the Polymyxins, which are antibacterial and 2) the Polyenes, which are primarily used against fungi and yeasts. Due to this host toxicity, these are used only in cases of Gm-ve bacteria resistant to multiple antibiotics. The Polyenes are a group of large, structurally related compounds that have no antibacterial activity, except some members of the genus *Mycoplasma*, but are highly active against yeasts and fungi. Polyenes act on sensitive fungi by binding to sterols of the plasma membrane resulting in alteration of membrane permeability and leads to the leakage of essential metabolites and eventually cell death.

Large Polyenes such as amphotericin B and nystatin exhibits 2 types of sterol binding, separable into fungistatic and fungicidal. The first is reversible and is associated with inhibition of growth and increased permeability of the membrane resulting in the loss of potassium from the cell. The second occurs at high drug concentrations, is irreversible and leads to cell death.

Mammalian cells due to the presence of cholesterol in their membranes are also subject to the action of Polyenes. This accounts for the very toxic and numerous side effects

associated with systemic polyene therapy such as headache, fever, chills, muscle and joint pain, nausea, vomiting, nephrotoxicity, neurotoxicity, hepatotoxicity, allergic reactions, etc.

Parenteral administration is only indicated in the treatment of serious infections. Amphotericin B is available as a lotion, cream or ointment for the treatment of cutaneous candidal infections and as lozenges for the treatment of oral candidiasis. Nystatin paint or lozenges can also be used for local application against candidal infections.

Interference with Nucleic Acid Synthesis

Several antimicrobial agents preferentially inhibit either DNA or RNA synthesis and most of them do not exhibit selective toxicity. However, nitroimidazole and quinolones do exhibit selective toxicity and are clinically useful in the treatment of certain bacterial infections.

Nitroimidazoles

- *Drugs include*—members of the nitroimidazole group of antibiotics include Metronidazole, Nimorazole, Tinidazole, Secnidazole and Ornidazole.
- *Mechanism of action*—the mechanism of action of these antibiotics is not clear. Since the drugs are only active against anaerobic organisms, it was first postulated that the drug interacted with biochemical pathways are found only in obligate anaerobes. This theory has been replaced, since current evidence indicates that these drugs interact with bacterial DNA. Upon entry into an anaerobic organism, metronidazole is reduced at the 5-nitro position producing a number of short-lived cytotoxic intermediates which react with DNA, resulting in cell death. The reductive pathways are characteristic of sensitive anaerobic bacteria and protozoa but are not present in aerobic bacteria. The reported carcinogenicity of this drug in rodents may be related to the fact that mammalian liver microsomes can reduce metronidazole.
- *Use in dentistry*—its primary use is in the treatment of obligate anaerobes associated with the oral cavity, intestinal and female genital tracts. Metronidazole has been used successfully in the treatment of periodontitis which is probably related to its high activity against the Gm-ve anaerobic bacilli that are often associated with the disease. The concurrent oral administration of metronidazole with either amoxicillin or Augmentin has been used with some success in the treatment of both juvenile and adult forms of periodontitis and also effective in the treatment of *A. actinomycetemcomitans*—associated periodontitis.

- *Absorption, fate and excretion*—metronidazole is rapidly and almost completely (80%) absorbed from the small bowel and very little reaches the colon. Food does not affect its bioavailability. The drug is metabolized mainly in liver. The metabolites are excreted mainly in the urine, imparting a deep red-brown color to it. The drug crosses the placental barrier and is present in the milk.
- *Adverse reaction*—nausea, anorexia, abdominal pain and metallic taste in mouth are common. Concurrent consumption of alcohol along with metronidazole causes antabuse like reactions due to accumulation of acetaldehyde in the blood. Metronidazole affects the activity of hepatic enzymes involved with the metabolism of ethanol and acetaldehyde. Possible tumorigenic effects are seen in experimental animals but not in clinical practice (But it should not be given to pregnant women or nursing mothers). The most severe, but rare reactions have been epileptic seizures and peripheral neuropathy.
- *Drug interaction*—alcoholic beverages should not be consumed during therapy with metronidazole for at least one day afterward, since abdominal cramps, nausea, vomiting, headache and flushing may occur. Toxic psychosis has been described when the drug has been administered to alcoholics receiving disulfiram within the last two weeks due to the possibility of psychotic reactions. Metronidazole potentiates the anticoagulant effect of warfarin and other oral coumarin anticoagulants.
- *Dosage*—Metronidazole (tablets—200/ 400 mg tid, IV—500 mg tid, Tinidazole, Secnidazole and Ornidazole have got longer half life (Dose: 500 mg bid for periodontal infections).

Quinolones

- *Drugs include*—the quinolones constitute a group of 1, 8-naphthyridine derivatives. These are synthetically produced drugs and therefore are not true antibiotics. Nalidixic acid was introduced into clinical use in 1964 and is the prototype of the quinolones drugs. Ciprofloxacin, a fluorinated quinolones carboxylic derivative is an example of the newer drug/2nd generation drug. Others in this group include Norfloxacin, Ofloxacin, Pefloxacin, Amifloxacin, Lomfloxacin, Levofloxacin, and Gatifloxacin.
- *Mechanism of action*—Nalidixic acid and Ciprofloxacin are generally considered bactericidal, since they inhibit the bacterial DNA replication (The bactericidal effect can only occur in the presence of competent RNA and protein synthesis. The imbalance of inhibited DNA replication and continuing protein synthesis results in inhibition of cell division). The site of inhibition is a

specific enzyme, DNA gyrase. This enzyme is responsible for the coiling of the long bacterial chromosome so that it fits into the bacterial cell, but still maintains its spatial arrangement. Quinolones interfere with this gyrase so that the spatial arrangement of the DNA is not possible and therefore it cannot be transcribed.

- **Ciprofloxacin**—ciprofloxacin is the 1st oral broad-spectrum antimicrobial agent with good activity against *Pseudomonas aeruginosa*. It has an excellent activity against a wide range of Gm-ve organisms, including many that are resistant to 3rd generation cephalosporins, broad-spectrum penicillins and the newer aminoglycosides. It also has good activity against Gm+ve bacteria and is thus useful in the management of mixed infections. However, most anaerobic bacteria are resistant. The newer quinolones may not be used routinely by dental professionals due to resistance encountered with the oral flora. Fortunately ciprofloxacin does not cross-react with most antibiotics. Infact, an additive effect has been reported with some antibiotics such as the β lactams, aminoglycosides, clindamycin and metronidazole.
- **Adverse reaction**—in comparison to other antibiotics, most quinolones are well tolerated. Nausea, headache, vomiting, diarrhea, photosensitivity, skin rashes, etc. are rarely seen. Ciprofloxacin is excreted in breast milk and may cross the placental barrier. Therefore, it should not be given to pregnant women or nursing mothers except in the case of severe infections. The quinolones are not recommended for pre-pubertal children.
- **Drug interaction**—quinolones inhibit the metabolism of theophylline and caffeine. Toxicity may occur due to the inhibitory effect of ciprofloxacin on theophylline if given together. Concurrent administration with antacids containing magnesium, Al, Ca, Zn containing multivitamins or iron containing preparations may substantially interfere with absorption of ciprofloxacin. Quinolones enhance the effect of warfarin and other oral *anti-coagulants*. It must be remembered that organisms do develop resistance to them and also since it is costly, their routine use in all infections is to be discouraged.
- **Dosage**—Ciprofloxacin (250/ 500 mg bid orally, 200/ 400 mg IV bid over 30-60 minutes), Ofloxacin (200-400 mg OD orally in the morning preferentially, 200-400 mg IV bid/ OD), Lomefloxacin (400 mg OD orally, 400 mg IV), Pefloxacin (400 mg bid orally, 400 mg IV bid over 1 hour), Sparfloxacin (200-400 mg OD/ bid orally).

Topical Antiseptic and Antibiotics

There are sufficient data to consider the species *Actinobacillus actinomycetemcomitans* (Aa), *Tannerella*

forsythensis and *Porphyromonas gingivalis* as key periodontal pathogens, whereas bacteria like *Prevotella intermedia*, *Campylobacter rectus*, *Peptostreptococcus micros*, *Fusobacterium nucleatum*, *Eubacterium nodatum*, *Streptococcus intermedia* and *Spirochetes* have shown moderate evidence for causation. Recent findings have associated yeasts, staphylococci, enterococci, pseudomonas, various enteric rods or some herpes viruses (Cytomegalovirus and Epstein-Barr virus type I) with destructive periodontal disease.

The clinical improvement after periodontal therapy indeed correlates directly with the degree to which pathogenic subgingival species are reduced or eradicated. To achieve reduction or elimination of pathogens, a subgingival application of antiseptics or antibiotics has often been considered.

Classification of Local Antimicrobial Therapy

Systemic antimicrobial agents enter periodontal pockets following their intestinal absorption and passage from the bloodstream into oral tissues, GCF and saliva. Systemic delivery provides a ready exposure of all periodontal sites to the antimicrobial agent, but it also poses a risk of adverse reactions to non oral body sites. Local antimicrobial therapy in periodontitis involves direct placement of an antimicrobial agent into subgingival sites, minimizing the impact of the agent on non-oral body sites. They are applied as a part of home oral hygiene regimens and professionally applied as part of office based treatment procedures. Local delivery may be further classified as providing either non-sustained or sustained subgingival drug delivery. Sub-gingival irrigation with antiseptic agents lacking substantively of oral tissues (Povidone-iodine) are examples of non-sustained drug delivery. Sustained or controlled drug release can be provided with sub gingival irrigation of agents intrinsically substantive for tooth root surfaces (aqueous tetracycline) or pocket placement of commercial antimicrobial fibers, gels or films.

- **Personally applied (Home self-care)**—Non-sustained subgingival drug delivery (Home oral irrigation) and sustained subgingival drug delivery (None developed to date).
- **Professionally applied (Dental office)**—Non-sustained subgingival drug delivery (Professional pocket irrigation) and Sustained subgingival drug delivery (Controlled-release device)

Agents that exert bactericidal effects within 5 minute time period are preferable for sub-gingival irrigation. Iodine showed 5 minute bactericidal action against the test organisms at concentrations therapeutically attainable at

Summary chart of antibiotics which are most commonly used in dentistry

Drug	Trade names	Dosage form	Dose	Indications	Contraindications	Adverse effects
Tetracycline	Achromycin, Terramycin	Capsule Injection Ointment	1-2 gm daily divided into 4 doses	Juvenile periodontitis, Chronic periodontal disease, Refractory periodontitis, Desquamative gingivitis	Pregnancy, children below 8 years	GI disturbance, tooth discoloration, enamel hypoplasia
Clindamycin	Clincin Dalcap	Tablet Capsule Injection	150-300 mg 6 hourly	Anaerobic streptococci, Clostridium infections, Refractory periodontitis	Hypersensitivity, infancy	Urticaria, diarrhea, abdominal pain, jaundice, liver abnormalities
Erythromycin	Althrocin Erythrocin E. mycin	Tablet	250-500 mg 8 hourly	Refractory periodontitis	Hypersensitivity	Mild epigastric pain, diarrhea, allergic reaction
Metronidazole	Flagyl Metrogyl Aldezole	Tablet Suspension injection	200-400 mg TDS. 200 mg/5 ml	Ulcerative gingivitis, periodontitis, ANUG	Neurological disease, blood dyscrasias, pregnancy	Anorexia, nausea, metallic taste, glossitis
Quinolone Ciprofloxacin	Ciplox Cipin	Tablet, injection	250-500 mg 12 hourly	Osteomyelitis, ANUG, joint infection, recurrent periodontitis	Hypersensitivity, children and elderly	Diarrhea, vomiting, abdominal pain, arthritis, restlessness
Penicillin Amoxicillin Ampicillin	Kaypen Ampilin Amox Mox	Capsule, Syrup	250 mg-1 gm TDS 125 mg/ml	Refractory periodontitis	Sensitivity Pregnancy	Skin rash, urticaria, GIT upset, diarrhea
Cephalosporin Cefotaxime Cefaclor Cefixime	Alcephin Biotax,	Capsule Injection Syrup	2-6 gm daily in 2-3 divided dose 4 gm initially followed by 2 gm IV	Inhibits growth of gram—ve anaerobes, Osteomyelitis	Hypersensitivity, renal failure	GIT upset, allergic reaction, increased SGPT, candidiasis
Chlorhexidine	Hexidine Rexitin	Suspension	0.2% w/v 100 ml	Oral hygiene bacterial infection	Allergy to chlorhexidine	Burning sensation, candidiasis, bad taste sensation, staining of teeth

subgingival sites (0.25-0.5%). But chlorhexidine needs (0.5-2%) and stannous fluoride needs (0.5-20%) 5 minutes for bacterial action. Metronidazole and amoxicillin required microbial contact time of 60 minutes or more for bactericidal activity. Interestingly, the bactericidal effects of povidone-iodine and of bicarbonate are potentiated by H_2O_2 . Chlorhexidine may be inactivated in periodontal pockets via binding to serum proteins, which are markedly elevated in GCF.

The other topical antimicrobial agents are : 1.64% SnF_2 gel, 3% H_2O_2 , 1% Chloramine-T, 1.25% amine fluoride gel, sodium bicarbonate tooth powder, and povidone-iodine etc.

Dose Calculation

Children Dose can be calculated by following formulas

$$\text{Young's Formula} = \frac{\text{Age} \times \text{Adult Dose}}{\text{Age} + 12}$$

$$\text{Clarke's Formula} = \frac{\text{Body Wt (kg)} \times \text{Adult Dose}}{150}$$

Prophylactic Antibiotics

According to AHA (American Heart Association, 2002)

- *Orally*—Amoxicillin 2 gm one hour before surgery. Children—50 mg/kg body wt. one hour before surgery.
- *IV or IM*—Ampicillin 2 gm half an hour before surgery. Children—50 mg/kg body wt. half an hour before surgery.
- *If allergic to penicillin*—Clindamycin 600 mg, children—50 mg/kg body wt. If Orally one hour before surgery, If IV or IM half an hour before surgery.

Others	Adult	Children
Clarithromycin	500 mg	15 mg/kg body wt.
Azithromycin	500 mg	15 mg/kg body wt.

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Analgesic and Anti-inflammatory Drugs

Introduction

In dentistry most common complaint of the patient is pain. So management of pain with the analgesic is primary importance of dentist. Pain is ill-defined, unpleasant sensation usually evoked by external or internal noxious stimulus. Pain receptors are mainly the free nerve ending widely spread in superficial layer and deep into the body. Certain pain receptors are present in the body called as chemoreceptor like bradykinin, prostaglandin (PG), prostacyclin (PGI₂), thromboxane (TXA₂), histamine, acetylcholine and proteolytic enzymes elicit pain after tissue injury. A delta fiber transmits fast-acute pain whereas c-fibers transmit slow-chronic type of pain.

Definition of Analgesic

Analgesics are the drugs that selectively relieve pain by acting in CNS or on peripheral pain mechanisms, without significantly altering consciousness. Transmitters involved in analgesic system are enkephalin and serotonin.

Classification (Fig. 41-1)

Nonselective COX inhibitors (Conventional NSAIDs)

- *Salicylates*—Aspirin, Calcium carbaspirin, Choline salicylates.
- *Propionic acid derivatives*—Ibuprofen, Naproxen, Ketoprofen, Flurbiprofen, Oxaprozin.
- *Anthranilic acid derivative*—Mephenamic acid.
- *Aryl acetic acid derivative*—Diclofenac.
- *Oxicam derivative*—Piroxicam, Tenoxicam.
- *Pyrrolopyrrole derivatives*—Ketorolac.
- *Indole derivative*—Indomethacin, Sulindac, Tolmetin, Etodolac.

- *Pyrazolone derivatives*—Phenylbutazone, Oxyphenbutazone.

Preferential COX-2 inhibitors

- Nimesulide, Meloxicam, Nabumetone.

Selective COX-2 inhibitors

- Celecoxib, Rofecoxib, Valdecoxib, Etoricoxib.

Analgesic-antipyretics with poor anti-inflammatory action

- *Para-aminophenol derivative*—Paracetamol (Acetaminophen)
- *Pyrazolone derivative*—Metamizol, Propiphenazone.
- *Benzoxazocine derivative*—Nefopam.

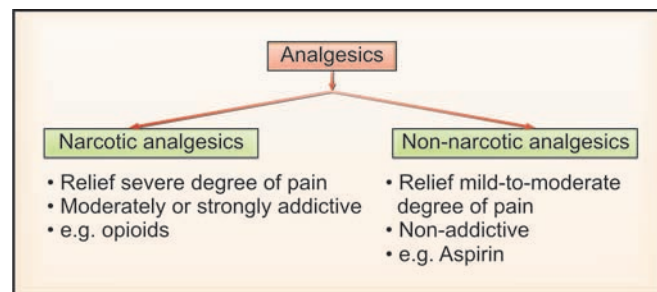


Fig. 41-1: Types of analgesics

General Principles of Use of Analgesics

- *Treat the cause of pain*—where possible, identify and treat the cause of pain.
- *Psychological management*—relieve the factors which lower the pain threshold (fatigue, anxiety and depression).
- *Regular analgesia*—chronic pain requires regular analgesia.
- *Simple analgesics*—use simple analgesics initially.
- *Do not use multiple drug*—avoid polypharmacy.

Nonselective Cyclo-oxygenase Inhibitors/Nonsteroidal Anti-inflammatory Drugs

NSAIDs mainly block the PG generation. PG is the substance which is produced from arachidonic acid by the enzyme cyclo-oxygenase (COX) which exist in a constitutive (COX-1) found in blood vessels, stomach and kidney; and an inducible form which is induced at the site of inflammation by cytokines and other inflammatory mediators. *Prostaglandin* play a part in the erythema, edema, and fever associated with inflammation and generate the pain either by its direct action or by sensitizing the pain receptors or by release of substance by tissue damage such as bradykinin and histamine. NSAIDs inhibit the action of cyclo-oxygenase (COX-1 and COX-2) enzyme. It interfere with the synthesis of prostaglandin and hence, causes the blockage of impulse generation mediating pain.

Nabumetone, a new NSAIDs preferentially inhibit COX-2. Similarly, newer drugs like Nimesulide and Meloxicam are also reported to have greater affinity for COX-2 as compared to COX-1.

Action of NSAIDs

- **Analgesic action**—NSAIDs do not affect the tenderness which is induced by direct application of PG, but it blocks the pain sensitizing mechanism induced by bradykinin, tumor necrosis factor Alfa, interleukins and other algesic factors. It is used in dental caries and postextraction pain.
- **Antipyretic action**—fever during infection is produced due to generation of pyrogens which induces the PG production. Hence, NSAIDs are used to block the action of pyrogens and help to reduce the body temperature in fever.
- **Anti-inflammatory**—PG are only one mediator of the inflammation. NSAIDs is considered to be inhibition of PG synthesis at the site of injury. Anti-inflammatory potency of different compounds correspond with their potency to inhibit COX. However, Nimesulide is a potent anti-inflammatory but weak COX inhibitors.
- **Dysmenorrhea**—PG synthesis in endometrium causes uncoordinated uterine contraction with compressed blood vessel results in uterine ischemia and pain. NSAID decrease PG synthesis and relieves the pain approximately 70%. It also relieves additional symptoms like headache, muscle ache and nausea.
- **Antiplatelet Function/Bleeding**—Platelet aggregation is due to TXA₂. NSAIDs inhibit TXA₂. NSAIDs acetylates COX present in platelets when they are in portal circulation, before it get acetylated in liver.

- **Closure of ductus arteriosus**—PGE₂ and PGI₂ produced in ductus arteriosus, kept it patent during fetal life. At the time of birth, it get closed as in the normal mechanism. But in case if it is failed to close during birth, small dose of indomethacin or aspirin induce its closure by inhibiting PG. But it should be avoided in pregnancy, as it may cause premature closure. So indomethacin and Aspirin is contraindicated during pregnancy.
- **Gastric mucosal damage**—Cyclo-oxygenase-1 (COX-1) has physiological role like protection of gastric mucosa by secretion of mucous. NSAID other than selective COX-2 inhibitor which may inhibit COX-1 cause gastric irritation and gastric ulceration due to reduced mucous and HCO₃ secretion. They induce back diffusion of H⁺ ions in gastric mucosa also. Paracetamol has weak inhibition of COX and selective COX-2 inhibitor do not cause gastric mucosal damage.
- **Renal effect**: COX-1 dependent impairment of renal blood flow and reduction. COX dependent Na and H₂O retention. Renal efferent is more significant in CHF, hypovolemia, hepatic cirrhosis.
- **Anaphylactic reaction**—aspirin precipitated asthma, angioneurotic swelling and urticaria or rhinitis in some individuals. Asthma is due to imbalance between constrictor PG (PGD₂), TXA₂, LT and dilators PG (PGE₂, PGI₂).

Salicylates

Aspirin

Aspirin is acetylsalicylic acid. It is rapidly deacetylated in body by acetylating other macromolecule like COX in platelet, before platelet get deacetylated in liver as physiologically happened. Acetylated platelet reduce tendency to aggregate and induce bleeding tendency due to aspirin. It is developed by Bayer in 1899. This includes species of willow. Salicylates derived from salix, a Latin name willow tree.

Mechanism of Action for Therapeutic Use (Fig. 41-2)

It acts both centrally but peripheral effect is main. It has therapeutic action of analgesic and antipyretic at low dose. At high dose anti-inflammatory action.

- **Analgesic and anti-inflammatory action**—its analgesic and anti-inflammatory action is due to prevention of PG (mainly PGE₂) mediated activated nerve ending. PG is associated with pain and fever related to inflammation and activates receptors released by tissue damage like bradykinin and histamine. They are chemical pain mediators. Aspirin reduce PG secretion, so it has

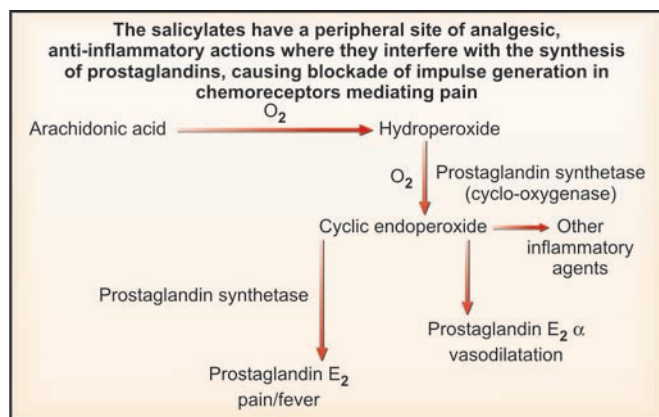


Fig. 41-2: Mode of action of non-narcotic analgesics

antipyretic and analgesic effect by inhibiting COX pathway by acetylating active site of COX and hence PG and TXA_2 . Its action is irreversible. It also blocks peripheral pain impulse generation. It has same action on subcortical site like thalamus and hypothalamus. Bradykinin and histamine brings inflammatory action during tissue damage (e.g. pulpitis), these chemicals are inhibited by Aspirin, and so it explains its anti-inflammatory action. Dose: Analgesic dose- 0.3-0.6gm for headache, toothache, joint pain, dysmenorrhea. Anti-inflammatory dose—3-5 g/day.

- **Antirheumatic action**—Bradykinin and prostaglandin cause vasodilatation, increase in capillary permeability which is sign of inflammation and inflammatory changes. These chemical mediators are inhibited by Aspirin which is demonstrated as an antirheumatic agent. It also prevents the release of lysosome which destroys cartilage of rheumatic joints. Dose- 3-5 g/day
- **Antipyretic action**—temperature regulating center present at hypothalamus during infection pyrogens directly act on hypothalamic integrating center (thermostat) causing fever. PGE_2 is also involved in this process. Aspirin reduces temperature by profuse sweating and synthesis of PGE in hypothalamus.
- **Antiplatelet effect**—therapeutically used in cerebral or myocardial infarction. Action of platelet is due to thromboxane produced during cyclooxygenase pathway in nucleated platelets causing aggregation of platelets in constricted vessel wall. Aspirin has antiplatelet action as it causes acetylation of COX in nucleus of platelet and reduces thromboxane synthesis. Hence non-nucleated, acetylated platelet irreversibly loses its aggregatory properties. Dose: 60-100 mg/day.
- **Other uses**—After birth, if ductus arteriosus is kept patent due to elaboration of prostaglandin. So, NSAIDs bring about closure of ductus within a few hours by inhibiting PG production.

Uses

- **As an analgesic**—for headache, backache, myalgia, joint pain, toothache, neuralgia, in low dose of 0.3-0.6 gm, 6-8 hourly.
- **As an antipyretic**—effective in the any kind of fever.
- **Acute rheumatic fever**—it is the first drug to be used in the dose of 4-6 gm or 75-100 mg/kg/day. It brings out marked symptomatic relief in 1-3 days and maintenance dose of 50 mg/kg/day continued for 2-3 weeks until all the signs of active disease disappear. Withdrawals of drug should be gradual over the next 2 weeks.
- **Rheumatoid arthritis**—Aspirin in the dose of 3-6 gm/day is effective producing relief of pain, swelling and morning stiffness.
- **Osteoarthritis**—it affords symptomatic relief only. It is used only when required.
- **Postmyocardial infarction**—as Aspirin inhibits the platelet aggregation, it lowers the incidence of reinfarction. It is used in the dose of 100-300 mg Aspirin/day for 12 weeks, reduces the transient ischemic attacks.
- Patent ductus arteriosus.

Adverse Effects

- **Peptic ulceration**—COX-1 has physiological role in gastrointestinal tract like mucus secretion for protection of gastric mucosa. Aspirin is COX inhibitor, it decreases mucus secretion from gastric mucosa by inhibiting cyclooxygenase. Treatment: Misoprostol (PG analog) is used to heal these ulcers.
- **Precipitation of asthma**— PGD_2 , TXA_2 are bronchoconstrictors while PGE_2 and PGI_2 are dilators. Aspirin causes imbalance between these constrictors and dilators causing precipitation of asthma.
- **Hemorrhage**—treatment is vitamin K supplements.
- **Salicylism**—large dose (3-5 g/day is anti-inflammatory dose) results in headache, dizziness, tinnitus, confusion, vertigo, reversible impairment of hearing and vision, sweating, palpitation and hyperventilation. These syndromes are known as salicylism.
- **Reye's syndrome**—use of Aspirin in children below 12 years with influenza or chickenpox cause life-threatening condition characterized by vomiting, lethargy, delirium, coma and even death. If a person survives, it causes irreversible brain damage. Use of Aspirin in children is prohibited in India.
- **Acute salicylates poisoning**—10-30 g is the suicidal dose of Aspirin in adults. In children, 4 ml is the fetal dose. Toxicity is seen with serum salicylates level more than 50 mg/dl.
- **Clinical feature**—vomiting, dehydration, electrolyte imbalance, acidotic breathing, abdominal pain, anorexia. In severe cases, CNS abnormalities and skin eruption, hyperpyrexia, restlessness, vertigo, tremors,

hallucination, convulsion and coma and even death due to respiratory failure and cardiovascular collapse may occur. Severe hypoglycemia may be seen in children.

- *Treatment*—gastric lavage to remove unabsorbed drugs, alkalization, diuresis. Activated charcoal is used to reduce absorption of salicylates from gut alcohol and adequate amount of intravenous fluid for dehydration. Bicarbonate solutions for alkaline diuresis. Glucose IV for ketosis and hypoglycemia. Blood transfusion or vitamin K for hemorrhage.

Contraindication

- *Peptic ulcer*—in peptic ulcers, bleeding tendencies.
- *Pregnancy*—in pregnancy, it causes premature closure of ductus arteriosus, delayed and prolonged labor, greater postpartum blood loss, low birth weight babies.
- *Nursing mother*—it is not used in breastfeeding mothers.
- *G6PD deficiency*—it causes hemolysis.
- *Before dental extraction*—it should be stopped 1 week before dental extraction or any major surgeries.
- *Chronic liver disease*—it may cause hepatic necrosis.
- *Diabetes*—Aspirin has hypoglycemic action.
- *Hepatic damage*—in hepatic damage, vitamin K deficiency or hemophilia, hypoprothrombinemia.
- *Chicken pox*—in children suffering from chicken pox or influenza—due to risk of Reye's syndrome in pediatric patient.

Pharmacokinetics

Aspirin absorbed from stomach and small intestine. Approximately 80% plasma protein binding capability. Half life at therapeutic dose is 15-20 mins, at anti-inflammatory dose is 8-12 hours and during poisoning 30 hours.

Drug Interaction

- It increases toxicity of oral anticoagulants, heparin, naproxen, oral antidiabetics, phenytoin, thiopental, indomethacin, valproic acid, and methotrexate.
- It decreases action of probenecid and sulfinpyrazone (uricosuric action)
- Ascorbic acid and ammonium chloride increase action of Aspirin
- Sodium bicarbonate decreases action of Aspirin
- It increases action of antiplatelet drugs
- Phenobarbital decreases aspirin action
- Aspirin decreases beta blocker's action
- In patient taking Aspirin, gastrointestinal bleeding increases by steroids, alcohol, indomethacin, and pyrazolon and anti-inflammatory drug. Furosamide increases Aspirin precipitation by decreasing salicylates excretion.

- Aspirin increases insulin and sulfonylureas.
- Antacid and activated charcoal decrease Aspirin absorption.

Trade Name

T. Aspirin 350 mg, T. Ecosprin 75, 150, 325 mg, T. Disprin 35 mg, T. Loprin 75 mg.

Propionic Acid Derivative

Ibuprofen

- *Action*—analgesic, antipyretic, anti-inflammatory.
- *Use*—analgesic (Dose—400 mg/TDS), antipyretic, anti-inflammatory. It is used in rheumatoid arthritis, osteoarthritis, ankylosing spondylitis, dental inflammation and other musculoskeletal disorders.
- *Adverse effects*—they are less than Aspirin. Gastric discomfort, nausea and vomiting. Headache, dizziness, blurring of vision, tinnitus and depression may occur. It precipitates aspirin-induced asthma.
- *Contraindication*—bronchial asthma, pregnant and lactating women, and peptic ulcer.
- *Pharmacokinetics*—99% bound to plasma protein, $t_{1/2}$ is 2 hours and it can cross blood-brain barrier, placenta.
- *Drug interaction*—it aggravates action of Aspirin, antiplatelet agent. It decreases action of diuretic and anti-hypertensive, e.g. beta blocker, thiazide.
- *Preparation*—T. Brufen 400, 200, 600, 800 mg. Ibugesic 100 mg/5 ml suspension.

Naproxen

- *Plasma $t_{1/2}$* —12-15 hours
- *Dose*—250 mg BID after meal
- *Trade name*—T. Naprosyn 500 mg, T. xenobid 250 mg.

Flurbiprofen

- *Action*—it suppresses both PGE₂ and PGE₂ alpha by cyclo-oxygenase enzyme. It inhibits leukocyte migration, 99% plasma protein binding.
- *Use*—as an anti-inflammatory in ocular infections—OCUFLUR (0.03%) eyedrop, 1 drop 6 hourly.
- *Trade name*—T. Arflur 50, 100 mg, T. Flurofen 100 mg. Ocuflur 0.03% eyedrop.

Anthranilic Acid

Mephenamic Acid

- *Action*—analgesic, antipyretic, anti-inflammatory in action.
- *Use*—as an analgesic primarily in muscles, joints and soft tissue pain. It is quite effective in dysmenorrhea. It may be useful in rheumatoid arthritis and osteoarthritis.

- *Pharmacokinetic*—Plasma $t_{1/2}$ 2-4 hours, absorbed slowly from gut.
- *Dose*—500 mg TDS
- *Adverse effect*—diarrhea, dizziness, rashes, hemolytic anemia, agranulocytosis, thrombocytopenic purpura, megaloblastic anemia.
- *Contraindication*—gastrointestinal ulceration, impairment of renal function.
- *Trade name*—C. Medol 250, 500 mg, T. Meftal 250, 500 mg, T. Ponstan 250 mg.

Aryl-Acetic Acid Derivative

Diclofenac

- *Action*—as an inhibitor of COX, it is analgesic, antipyretic, anti-inflammatory drug.
- *Plasma half-life*—2 hrs.
- *Indication*—rheumatoid arthritis, osteoarthritis, bursitis, post-traumatic and post-operative inflammation and ankylosing spondylitis.
- *Dose*—100-200 mg BID or TDS.
- *Trade name*—T. Diclomax 25, 50, 100 mg; Voveran IM 75 mg; T. Voveran 50 mg.
- *Adverse effect*—epigastric pain, nausea, headache, dizziness, rashes. Kidney damage is rare.

Oxicam Derivatives

Piroxicam

- *Action*—it reversibly inhibits COX, so it has analgesic, antipyretic, anti-inflammatory properties.
- *Plasma half-life*—2 days
- *Adverse effect*—heart burn, nausea, anorexia.
- *Indication*—rheumatoid and osteoarthritis, ankylosing spondylitis, postextraction pain, pulpitis in dentistry.
- *Dose*—20 mg BD for 2 days followed by 20 mg OD.
- *Trade name*—C. Pirox 10, 20 mg, C. Pirox 10, 20 mg. Tenoxicam: T. Tabitil 20 mg Dose: 20 mg OD. T. Dolonex.

Pyrrolo-pyrrole Derivative

Ketorolac

- *Action*—it also inhibits PG and relieves pain by peripheral mechanism.
- *Half-life*— $t_{1/2}$ 5-7 hour.
- *Indication*—Postoperative, dental and acute musculoskeletal pain, pain due to bony metastasis, migraine.
- *Adverse effect*—nausea, abdominal pain, ulceration, loose stools, drowsiness, pain at injection site, dizziness.
- *Dose*—Dose: 10-20 mg tablets 6 hourly. 15-30 mg IM or IV every 4-6 hours

- *Trade name*—T. Ketorol 10 mg, T. Toralac 10 mg, T. Ketanov 10 mg.

Preferential COX-2 Inhibitors

Nimesulide

- *Action*—it has weak inhibition of PG synthesis but action on COX-2 selectively, has potent anti-inflammatory action by inhibition of histamine release.
- *Use*—postoperative dental pain, dysmenorrhea, inflammation of the ear and throat.
- *Adverse effect*—nausea, loose motion, rash, pruritus, dizziness, hepatic failure.
- *Pharmacokinetic*—99% plasma protein binding
- *Plasma half life*—2-5 hours
- *Dose*—100 mg BID
- *Trade name*—T. Nimulid 100 BD, T. Nimegesic, T. Nimodol.

Para-amino Phenol Derivative

Paracetamol

- *Action*—it is de-ethylated active metabolite of phenacetin. It has analgesic-antipyretic action but no anti-inflammatory action. It has poor inhibition of PG, poor ability to inhibit COX in presence of peroxide which is synthesized at inflammation site. This peroxide is absent in brain, so it is more active on COX in brain (it has no effect on blood clotting mechanism).
- *Use*—headache, musculoskeletal pain, dysmenorrhea. It is first drug of choice for osteoarthritis and is also used in patient in whom Aspirin is contraindicated.
- *Pharmacokinetic*—it is well absorbed orally.
- *Plasma half life*—2 hours
- *Dose*—0.5-1 gm 6 hourly or TDS
- *Adverse effect*—it has safer action. On prolonged use liver damage can occur. Acute Paracetamol poisoning: Early manifestations are: nausea, vomiting, abdominal pain. After 12-18 hours, centrilobular hepatic necrosis and renal tubular necrosis, hyperglycemia and then coma. Treatment: Induce vomiting or gastric lavage. Activated charcoal to prevent further absorption.
- *Trade name*—T. Crocin 0.5, 1 gm, febrinil 300 mg/2 ml inj. T. Calpol 1 gm.

Patient Instruction for Use of Non-steroidal Anti-inflammatory Agents

- Take with a full glass of water.

- Take with food to minimize gastrointestinal irritation.
- Use caution with driving because of possible drowsiness or dizziness.
- If pain does not subside within a few days, call the dentist.

Opioid Analgesics

The dark brown, resinous material obtained from poppy capsule is called '*opium*'. These are plant products containing pharmacologically active ingredient i.e. alkaloid. It contains two types of alkaloids, viz. phenanthrene derivatives and benzoisoquinoline derivatives. They are also called *narcotic* drugs. The term narcotic is derived from the Greek word meaning '*stupor*' and was used for agents that produce sleep. The opioids bind to receptors located in the central nervous system, producing an altered perception of and response to pain.

Pharmacological Actions

- *Analgesia*—these drugs relieve severe pain like that of visceral pain and pain of trauma. Mechanisms by which they act are as follows:
 - They increase the threshold for painful stimuli. They act directly on the receptor site in the CNS to inhibit release of excitatory transmitters from primary afferents carrying pain impulses and elevate the pain perception threshold.
 - They alter emotional reaction to pain. They produce sleep which also elevates the pain threshold.
- *CNS*—these drugs have a site specific depressant and stimulant action in the CNS. It produces euphoria in the presence of pain. However, in absence of pain, it produces dysphoria; and, with an increased dose it produces sleep.
- *Respiration*—respiratory depression by reducing the sensitivity of medullary respiratory center neuron to CO_2 ; rate and tidal volume are both decreased.
- *Pupil*—miosis resulting in pin-point pupils.
- *CTZ*—nausea and vomiting can occur resulting from stimulation of the CTZ (chemoreceptor trigger zone) especially when the stomach is full.
- *Cough*—suppress cough by depressing the cough center.
- *Neuroendocrine*—the sex hormone and corticosteroid levels are lowered in the short term, but tolerance develops in the long term. Morphine can release ADH and reduce urine volume.
- *GIT*—these drugs increase tone and segmentation but decrease peristaltic propulsive movements. This produces spasm of intestine and sphincters. Secretion of HCl and intestinal secretions are decreased. These drugs increase absorption of H_2O , so lead to constipation.
- *Biliary tract*—there may be increase in intrabiliary pressure and may cause biliary colic.
- *CVS*—they cause vasodilatation by decreasing tone of blood vessels, histamine release and depression of vasomotor center. Hypotension may occur at toxic doses. Opioids are effective in relieving dyspnea of left ventricular failure because of reduced venous tone and decreased peripheral resistance.

Pharmacokinetics

- *Absorption*—Most opioid analgesics are absorbed well when taken orally. Absorption occurs from the lungs and from the nasal and oral mucosa whereas through GIT is slow and incomplete. A quick effect occurs following a subcutaneous injection.
- *Distribution*—they are partially bound to plasma proteins. The opioids can affect fetus in pregnant women.
- *Metabolism*—they are metabolized by conjugation with glucuronic acid in the liver.
- *Excretion*—they are excreted by glomerular filtration in the urine within 24 hours.

Adverse Effects

- *Constipation*—the opioids produce constipation by producing tonic contractions of the gastrointestinal tract.
- *Respiratory depression*—the opioid analgesic agonists depress the respiratory center in a dose-related manner. The depression is related to decrease in sensitivity of the brainstem to carbon dioxide.
- *Nausea and vomiting*—opioids often produce nausea and vomiting which is due to their direct stimulation of the chemoreceptor trigger zone (CTZ), located in the medulla.
- *Miosis*—the opioid analgesics cause miosis, an important sign (pinpoint pupils) in the diagnosis of an opioid overdose or an identical in addicts.
- *Urinary retention*—it increases smooth muscle tone in the urinary tract, thereby causing urinary retention.
- *CNS effects*—it may produce CNS stimulation exhibited by anxiety, restlessness, or nervousness.
- *Cardiovascular effects*—it may depress vasomotor center and stimulate the vagus nerve. With high dose, postural hypotension, bradycardia, and even syncope may result.
- *Biliary tract constriction*—in high doses, the opioids may constrict the biliary duct, resulting in biliary colic.
- *Apnea*—it may occur in newborns, when these drugs are given to mothers during labor pain.
- *Addiction*—it is proportional to their analgesic strength. As the use of opioids in dentistry is usually very less, this does not produce problems for dentist.
- *Allergic reaction*—it includes skin rashes and urticaria.

Summary chart						
Drug	Trade name	Dosage form	Dose	Indication	Contra-indication	Side effect
Salicylates (Aspirin)	Disprin Anacin Colsprin Biospirin	Tablet Injection	Analgesic / antipyretic— 0.3-0.6 gm TDS	Dental pain Headache Fever Arthritis Osteoarthritis Postmyocardial infarction	Hemophilia Hypersensitivity Peptic ulcer Gout Asthma Patient on anticoagulants	Nausea Vomiting GI disturbance Tinnitus Vertigo
Para-amino phenol derivative (Paracetamol)	Calpol Crocin Metacin Pacimol Febrinil	Tablet Injection Syrup Drops	0.5-1 gm TDS infant—50 mg 1-3 year— 60-80 mg 4-8 year— 240—320 mg 9-12 year— 300-600 mg	Pain Fever	No absolute contraindication Special precaution in hepatic and renal failure	Nausea Rash Leucopenia
Indole derivatives (Indomethacin)	Indocap Indoflam	Capsule Tablet Injection	25-50 mg 2 to 4 times a day neonates— 0.2 mg/ kg	Dental pain Rheumatoid arthritis Osteoarthritis Ankylosing spondylitis	Hypersensitivity Epilepsy Kidney disease Pregnancy Psychiatric cases	Gastric irritation Nausea Anorexia Diarrhea Asthma
Propionic acid derivatives (Ibuprofen Naproxen Ketoprofen)	Brufen Enflam Febrilix Combiflam Duoflam Nalyxan (Naproxen) Ostofen (Ketoprofen)	Tablet Capsule	Ibuprofen— 200-600 mg TDS Naproxen— 250 mg BD Ketoprofen— 50 mg BD	Postextraction pain Soft tissue injuries Rheumatoid arthritis Musculoskeletal disorders	Pregnancy Peptic ulcer Gastrointestinal bleeding Laceration	Gastrointestinal disorder Rash Hypersensitivity
Oxicam Piroxam	Pirox Dolonex Movon	Capsule Tablet Injection Gel	20-40 mg in divided or single doses	Post operative pain Osteoarthritis Rheumatoid arthritis	Hypersensitivity Pregnancy Lactation Children below 6 years Bronchial asthma	Heart burn Nausea Anorexia Rashes Pruritis Edema
Nimesulide	Nise Nulide Emsulide Nimsaid Nimulid Nimol	Tablet	Adult—100mg BD Children— 5 mg/kg /day in 2-3 divided doses	Dental pain Arthritis Gout Painful conditions Musculoskeletal pain	Peptic ulcer Hypersensitivity Hepatic impairment Pregnancy	Epigastric distress pain Nausea Rashes Edema
Aryl acetic acid derivatives Diclofenac sodium	Diclofen Diclomol Relaxyl Voveran	Tablet Gel	100-150 mg daily in 2-3 divided doses	Dental pain Arthritis Gout Painful post-operative condition	Peptic ulcer Hypersensitivity Pregnancy Lactation Asthma	Epigastric pain Nausea Rashes Edema Itching
Anthranilic acid Mephennamic acid	Mefac Meftal forte Spasmodol forte	Tablet Capsule Suspension	250-500 mg TDS	Pain Arthritis Osteoarthritis Dysmenorrhea As antipyretic	Hypersensitivity Epilepsy	Epigastric distress Rashes Hemolytic anemia Diarrhea
Ketorolac	Ketorol Ketanov Torolac	Tablet Injection	20 mg initially followed by 10 mg 4-6 hourly 30-60 mg IM followed by 10-30 mg 4-6 hourly	Short term management of postoperative pain Musculoskeletal pain	Hypersensitivity Syndrome of nasal polyps Peptic ulcer Coagulation disorders Angioedema	Gastric ulceration Bleeding Pain edema Headache Sweating

Drugs used in combination				
Drug	Dosage	Action and use	Adverse effect	Contraindication
Paracetamol + Ibuprofen	Adult— oral 325-500 mg Paracetamol + 100 mg Ibuprofen Children 125 mg Paracetamol + 100 mg Ibuprofen. TDS	It is a potent anti-inflammatory with analgesic and antipyretic action. Chronic pain, inflammatory dental conditions	Nausea, vomiting GIT and vision disturbances, rash	Active peptic ulcer, recent GI bleeding
Acetofenac + paracetamol	100 mg + 500 mg	It inhibits the synthesis of PGE ₂ and stimulates the synthesis of GAG. Chronic pain, inflammation	Nausea, vomiting, headache, vertigo, tinnitus	Hypersensitivity, pregnancy, peptic ulcer

- *Miscellaneous*—idiosyncrasy, allergy, tremor, delirium, sedation, mental clouding and lethargy.

Precaution and Contraindications

- *Infant and elderly persons*—they are more susceptible to respiratory depression.
- *Head injury*—as opioids depress the respiratory center, reduced ventilation produces vasodilation, and an increase in intracranial pressure.
- *Respiratory insufficiency*—it is dangerous to give these drugs in respiratory insufficiency as sudden death can occur.
- *Bronchial asthma*—morphine can precipitate an attack by its histamine releasing action.
- *Hypotensive states and hypovolemia*—there may be a fall in blood pressure.

Therapeutic Uses

- *Analgesics*—they are indicated in severe pain of any type. They provide symptomatic relief without affecting the cause. Opioids are used in traumatic, visceral, ischemic, postoperative, burn and cancer pain.
- *Preanesthetic medication*—morphine and pethidine are commonly used as preanesthetic medication.
- *Balanced anesthesia and neurolept analgesia*—morphine, pethidine and fentanyl are important components of these techniques.
- *Relief of anxiety and apprehension*—especially used in myocardial infarction and internal bleeding.
- *Acute left ventricular failure*—morphine affords dramatic relief by reducing preload on the heart due to

vasodilatation and peripheral pooling of blood.

- *Cough*—codeine or its substitutes are widely used for suppressing dry, irritating cough.
- *Diarrhea*—the constipating action of codeine has been used to check diarrhea and to increase consistency of stools.

Uses in Dentistry

- *Postoperative pain*—main use of narcotic analgesics in dentistry is for severe postoperative pain unresponsive to the anti-inflammatory analgesics.

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Introduction

Fungi are eukaryotes and lack chlorophyll pigment. It possesses differentiated nuclei surrounded by nuclear membrane and reproduce either by budding or forming spores. It has rigid chitinous cell wall. Morphologically it may be either oval cells or long tubular septate hyphae showing true lateral branching. Fungi can be mould type, yeast type, yeast-like fungi (*Candida albicans*) and dimorphic fungi (Fig. 42-1).

Antifungal drugs are used in the treatment of superficial (Affecting oral mucous membrane, skin, nails, etc) and deep fungal infection (deeper tissues and organs). Fungal infections are caused by broad spectrum species of yeast like fungus including *Candida albicans*, *Cryptococcus neoformans*, *Histoplasma capsulatum*, *Blastomyces dermatitidis*, *Coccidioides immitis*, *Aspergillus fumigatus*, and many other pathogenic fungi.

Among these, *Candida albicans* has become most common organisms which is isolated from blood culture causes mainly oral fungal infection called as oral candidiasis or oral thrush. Oral candidiasis is most common fungal infection caused in humans and as variety of clinical

manifestations, it may be associated with the use of broad spectrum antibiotics, corticosteroids, cytotoxic drugs indwelling catheters, immunosuppressive therapy, dental implants, emergence of AIDS.

Types of Fungi

Fungi are either commensals or pathogenic. Pathogenic produces mycotoxins and enzymes causing harm.

- *Pathogenic fungi:*
 - Superficial
 - Deep
 - Truly pathogenic.

Classification

First Classification

Antibiotics

Polyenes

- Amphotericin B
 - Nystatin
 - Hamycin
 - Natamycin
- ##### Heterocyclic benzofuran

- Griseofulvin

Anti-metabolites

- Flucytosine

Azoles

Imidazole

Topical

- Clotrimazole
- Econazole
- Miconazole

Systemic

- Ketoconazole



Fig. 42-1: Different types of fungi.

Triazoles**Systemic**

- Fluconazole
- Itraconazole

Allylamine

- Terbinafine

Other topical agents

- Tolnaftate
- Undecylenic acid
- Benzoic acid
- Cyclopiroxolamine
- Quiniodochlor
- Sodium thiosulfate

Second Classification

According to its mode of action:

- Drugs that disrupt the fungal cell membrane-
 - Polyenes: Amphotericin B
 - Azoles: Imidazole, e.g. Ketoconazole, triazoles, Fluconazole
 - Allylamine: Terbinafine
- Drugs that inhibit mitosis: Griseofulvin
- Drugs that inhibit DNA synthesis: Flucytosine

Nystatin

It is polyene antibiotic obtained from *Streptomyces noursei*.

- **Trade name**—MYCOSTATIN
- **Preparation available**—Oral—5,00,000 unit tablets; Topical-100,000 unit/gm cream and ointments, 100,000 units vaginal tablets. It is available currently for topical use in creams, ointment, suppositories, lotion, suspension, vaginal tablets, and oral pastille.
- **Mechanism of action**—depending upon concentration used, it can be either fungistatic or fungicidal agent. Ergosterol is sterol found in cell membrane of fungi. It binds to the ergosterol and increases the permeability of cell membrane facilitating pore formation. These micropore formation allow leakage of intracellular ions and macromolecule leading to cell death.
- **Indication**
 - **Oral candidiasis (Oral thrush)**—Nystatin is drug of choice. it is commonly used as topical agent to suppress Candida infection. It is not administered parentally as it is toxic for systemic use. (1) Thrush is treated by holding 5 ml of nystatin suspension (for infants- 2 ml) in oral cavity for several minutes 4 times daily before swallowing. (2) Alternative treatment is to retain vaginal tablet in the mouth until it dissolves, at least 4 times a day. (3) Nystatin oral pastille (200,000 units) dissolved slowly in the mouth 5 times a day—can be mixed with glycerin. (4) Mycostatin creams and ointments (1 lack units) may be used in

candidiasis with angular cheilitis, stomatitis. (5) Mycostatin cream and ointments with absorbable corticosteroids and antibiotics may accelerate symptomatic effect. (6) Mycostatin oral rinse—1 teaspoon of nystatin oral suspension (100,000 unit/cc) mixed with ¼ cup of water is used as oral rinse for 3-4 times a day for 7-10 days.

- **Denture stomatitis (chronic atrophic candidiasis)**—0.2% chlorhexidine solution with Mycostatin tablet dissolves in it forming Gel are used mainly in such patients. In denture wearing patients, nystatin ointments is to be applied to the fitting surface of clean denture thoroughly and regularly and should be left out of mouth at night keeping it in hypochlorite solution.
- **Candidial angular cheilitis**—treatment with nystatin or Mycostatin yield good result.
- **Monilial diarrhea and monilial vaginitis**—nystatin is drug of choice. It is highly effective in monilial vaginitis in the dose of 1 tab 3-4 times a day (For children 1-2 million unit/day in divided doses in every 4-6 hours). For topical application, dissolves 1 tab in 5 ml of glycerin and then use.
- **Recurrent perianal/vaginal/vulvar and diaper candidiasis**—it is used topically.
- **Vulvovaginal candidiasis**—it is treated by application of vaginal tablet twice a day for 14 days, then nightly for an additional 14-21 days.
- **Mucocutaneous candidiasis**—topical Nystatin agent is applied on skin and mucosal surface 4 times a day gives better results.
- **Corneal infection / uterine infections**—cream, ointment, and vaginal tablets are commonly used. Nystatin is well tolerated in eye at the dose of 1,00,000 unit .1) Topical treatment: Eye drops containing nystatin 5-10 microgram. 2) Systemic treatment: nystatin tablets (250 mg) TDS for 2 weeks followed by 1 troche per day for three weeks.
- **Adverse effect**—fever, chills, headache, hypotension, rashes, nausea, and vomiting.
- **Contraindication**—Hypersensitivity reaction.

Amphotericin-B

Amphotericin B is an antifungal antibiotic produced by *Streptomyces nodosus*.

- **Trade name**—Fungizon, Abelcet, Ambisome, Amhotec.
- **Preparation available**—Parenteral: conventional formulation: Fungizone 50 mg power for injection. Lipid formulation: Abelcet—100 mg/20 ml suspension for injection. Ambisome—50 mg powder for inj. Amphotec—50,100 mg powder for inj. Topical: 3% cream, lotion, ointment.

- **Mechanism of action**—it binds to the ergosterol, the sterol of cell membrane of fungi increases the permeability of cell membrane facilitating pore formation. These pore formation allow loss of cation, intracellular ions and loss of calcium leading to cell death. It acts as fungicidal at high concentration and fungistatic at low concentration. It is effective against numerous fungi and yeast like *Candidia albicans*, *Cryptococcus neoformans*, *Histoplasma capsulatum*, *Blastomyces dermatitis*, *Coccidioides immitis*, *Rhodotorula*, *Aspergillus*, *sporothrix*.
- **Indication**
 - **Candidiasis**—amphotericin B remains the drug of choice for mainly oral, vaginal and cutaneous candidiasis. It is given in oral candidiasis (Oral thrush), acute atrophic candidiasis (antibiotic stomatitis), denture sour mouth (chronic atrophic candidiasis), chronic hyperplastic candidiasis (candidial leukoplakia). 1) Amphotericin B is mostly applied topically in the form of ointment, suspension, drops, cream, and lotion. It is applied 2-3 times a day. 2) In oral candidiasis- *Mystelcin elixir* containing both tetracycline and amphotericin used topically may prove to be beneficial. 1 ml suspension (100 mg/20 ml) is vigorously swished in the mouth and then swallowed, 4-5 times for 2-1 weeks. 3) In denture wearing patient, amphotericin B ointment applied to fitting surface of denture before it use. In candidial keratitis / corneal ulcers (conjunctivitis)- It is mostly applied topically by eyedrops containing 2.5 to 10 mg/ml. It cures only superficial infection because of its poor penetration. Subconjunctival injections are quite painful. Amphotericin B contains deoxycholate sodium which is highly toxic and irritating to eyes. Large doses may cause tissue necrosis.
 - **Mucosal leishmaniasis**. It is treated with intravenous Amphotericin B 0.1 to 1 mg/kg on alternate day with total dose of 1.5-2 gm.
 - **Fungal meningitis/cryptococcal meningitis**—intrathecal therapy with Amphotericin B is highly successful.
 - **Systemic fungal disease**—250 mg/kg daily, gradually increased if tolerated to 1 mg/kg by slow intravenous infusion over 4-6 hrs. Max. 1.5 mg/kg daily or on alternate day to a total dose of 3-4 gm is given over 2-3 months.
 - **Intestinal moniliasis/aspergillosis**—It is effective on topical use. It can be administered systemically in the dose of 50-100 mg in tablet form.
 - **Paronyllosis and intertriginous candidiasis**—apply 2-4 times daily to the affected area.
 - **Coccidioidomycosis**—it is given in Coccidioidal granuloma, valley fever, desert fever. Long term systemic therapy is required for complete cure.
- **Chromomycosis, chromo-blastomycosis, Phaeohyphomycosis**—Amphotericin B, intravenously or intralesional yield good treatment result.
- **Cryptococcosis (torulosis, European blastomycosis)**—intraoral lesions present over hard palate, extraction socket, tongue are mainly treated by oral application/topical use of Amphotericin B cream, ointment, 4-5 times daily for at least 14 days.
- **Histoplasmosis**—treatment includes systemic and topical use of Amphotericin B is the drug of choice. It gives good result. Long-term therapy may require.
- **Mucormycosis/Sporotrichosis/blastomycosis**—amphotericin B can use as an alternative drugs.
- **Adverse effect**—it is toxic drug causes impairment of renal and hepatocellular function, anemia, mental and neurological changes, renal toxicity.
Chills, fever, vomiting, headache, thrombophlebitis may occur if injected IV.

Hamycin

It is polyene antibiotics obtained from *Streptomyces pimprina*.

- **Trade name**—HAMYCIN
- **Preparation available**—Topical -4 lac unit/gm ointment, 5 lac unit vaginal ovules. 2 lac/ml suspension.
- **Mechanism of action**—same as Nystatin.
- **Indication**—oral thrush (due to *Candida albicans*)—ointment or suspension is applied to the affected area of oral mucosa with sterile cotton 2-3 times a day for 7-10 days. It can also be given in cutaneous candidiasis, monilial or *Trichomonas vaginitis*, histoplasmosis, pulmonary mycosis and otomycosis caused by *Aspergillus*. Dose recommended is 10-20 mg/kg daily.
- **Adverse effect**—mild rashes, irritation.
- **Contraindication**—Hypersensitivity.

Natamycin

This is a polyene antibiotic topically effective antifungal agent.

- **Trade name**—NATAMYCIN, PIMAFUCIN
- **Preparation available**—Topical- 5% suspension, 2% cream, 1% ointment and 25 mg vaginal Tabs.
- **Mechanism of action**- same as Nystatin.
- **Indication**—
 - Oral thrush/ Oral candidiasis: Natamycin vaginal tab or ointment is applied on affected area of oral mucosa for several minutes 3-4 times daily.
 - It can be given in vaginal candidiasis (25 mg vaginal tab is used topically).
 - *Trichomonas vaginitis* (used only topically).

- Ocular keratitis caused by fusarium, cephalosporium and other fungi. 5% Natamycin ophthalmic suspension as eyedrop is used topically 3-4 times a day.
- *Adverse effect*—nausea, mild rashes occasionally.

Griseofulvin

It is extracted from penicillium Griseofulvin. Griseofulvin was the first orally effective antifungal agent. It is effective against dermatophytes including Epidermophyton, Trichophyton, microsporum, etc. It is ineffective against Candida and *P. orbiculare*.

- *Trade name*—Dermofulvin, Grisactin, Grifulvin.
- *Preparation available*—Oral microsize: 125, 250 mg capsules, 250, 500 mg tablets, 125 mg/5 ml suspension. Oral ultramicrosize: 125, 165, 250, 330 mg tab.
- *Mechanism of action*—Griseofulvin binds to keratin in new forming skin cells and protects the skin from new infection. So, its action is to prevent infection of this new skin structure. It does not kill fungus which are already present.
- *Indication*—
 - *Dermatophytic infections*—Griseofulvin is effective against all superficial ringworm infection (skin and hair infection). It must be applied for 2-6 weeks on affected skin and hair to allow replacement of infected keratin.
 - *Fungal infection of nails and scalp*—Long therapy of griseofulvin may require to allow regrowth of new protected nails. For adult, 500 mg daily in single or divided doses with meal; For children, 10 mg/kg of body wt daily in single or divided dose with meals. Oral suspension is also available. Fingernails may respond to 6 month of therapy whereas toe nails may require 8-18 months.
 - *Fungal infection of tinea of hand, athlete's foot, tinea pedis, tinea manus*—it may require long therapy 500 mg daily in single/divided dose with meals for 3-4 weeks.
- *Adverse effect*—mild rashes, nausea, vomiting and headache, gastrointestinal upset, photophobia, peripheral neuritis.
- *Contraindication*—porphyria, liver disease, systemic lupus erythematosus.

Flucytosin

It is oral antifungal agent effective against systemic infection due to yeast and fungi. Its spectrum of action is much narrower than that of Amphotericin B.

- *Trade name*—ANCOBON
- *Preparation available*—Oral: 250, 500 mg capsule. 100-150 mg/kg/day in patient with normal renal function.

- *Mechanism of action*—it has antifungal action. Susceptible fungal cell take up flucytosin, convert it to fluorouracil, which is converted to 5-fluorodeoxyuridylic acid which inhibit DNA and RNA synthesis. But human cells are unable to convert parent drug to its active metabolites, so it is relatively non-toxic. It is widely distributed in the tissue including blood and CSF.
- *Indication*—it is mainly used as an adjuvant drug. *Cryptococcal meningitis*—It is used in amphotericin B to reduce its toxicity in dose of 100 mg/kg/day in 4 divided doses by an oral route. Systemic candidiasis with uterine/GIT infections. Aspergillosis and chromoblastomycosis: Flucytosin is used in combination with itraconazole.
- *Adverse effect*—prolonged use with high doses cause leukopenia, thrombocytopenia, and anemia due to bone marrow depression, nausea, vomiting, and skin rashes.

Clotrimazole

It is used exclusively as topical agent. It is toxic on parental administration.

- *Trade name*—Mycoban, Lotrimin Mycelex, Surfaz.
- *Preparation available*—Topical: Mycoban Gel 12% (30 gm), Gynostatum vaginal tab, 1% vaginal cream, lotion, solution. Oral: 100 mg, 200 mg, 500 mg tab.
- *Mechanism of action*—it inhibits the synthesis of ergosterol by binding to catalytic haem ion of fungal cytochrome resulting in damage to cell membrane of fungi.
- *Indication*—
 - Candidiasis caused by *Candida albicans*, oral candidiasis, cutaneous candidiasis.
 - Treatment of tinea pedis (athlete's foot), tinea cruris (jock itch), tinea corporis (ringworm).
 - Vaginal infection—1 vaginal tab 100mg intra-vaginally at bed time for 7 consecutive days. Then in alternative days 1 tab (200 mg) intravaginally at bedtime for 3 consecutive days.
 - *Dermatophytic infections like candidiasis and seborrheic dermatitis*—Topical antifungal-corticosteroid fixed combination has been recently introduced. Clotrimazole-betamethasone dipropionate cream (lotrisone) is used once or twice daily, applied to the affected area results in the clearing of superficial dermatophytic infection in 2-3 weeks.
 - *Skin infection caused by corynebacteria*—it is well tolerated by topical use.
 - *Oropharyngeal candidiasis* : it is used in the dose of 10 mg daily (TDS)
 - *Pityriasis versicolor*—100 to 500 mg/daily is quite effective.

- Toxoplasmosis, Aspergillus infection of eye—1% suspension is to be applied topically hourly, then 4 times a day for 8-12 weeks. In case of intraocular infection, both topical and oral dosage of 60 mg/kg/day for 1-2 weeks to achieve normal intraocular level, then discontinue.
- *Adverse reaction*—local burning or stinging, skin irritation, rash, hypersensitivity.
- *Contraindication*—1st trimester of pregnancy.

Econazole

It is antifungal agent mainly used topically.

- *Trade name*—ECANOL, SPECTAZOLE.
- *Preparation available*—Topical: 1% cream-10 gm.
- *Mechanism of action*—same as Cotrimazole.
- *Indication*—
Treatment for trichomoniasis—1% ointment, 150 mg vaginal tablets is applied to affected area of skin or oral mucosa twice a day for 7-10 days. Mucocutaneous candidiasis—it is used as an oral rinse, cream, ointment as well as suspension. Vaginal tab dissolved in ¼ cup of water and used as oral rinse for oral candidiasis. Dermatophytosis—mostly applied topically 2-3 times a day. Pityriasis versicolor.
- *Adverse effect*—local burning or stinging, skin irritation, rash.

Miconazole

It is used mostly as topical antifungal agent. It is more toxic than ketoconazole.

- *Trade name*—MICOGEL, MONISTAT, MICATIN.
- *Preparation available*—Topical: ointment 2%-15 gm, lotion 2%-15 ml, powder 2%-15 gm, cream 2%-15 gm, gel 2%-15 gm, vaginal suppositories- 100, 200 mg.
- *Mechanism of action*—same as Cotrimazole.
- *Indication*—
 - *Severe systemic fungal infection*—in adult, 1200 to 2400 mg/day in divided doses.
 - *Cutaneous candidiasis*—applied cream, ointment to affected area twice daily for 7 days.
 - *Systemic mycosis*—Miconazole is used by intravenous route (Monistat).
 - *Ocular infection*—Topical (1%) for every hours and subconjunctival injection of 5 mg miconazole (1%) MICOPTIC OPICOPS once/twice a day.
 - *Other*—chronic mucocutaneous candidiasis, fungal meningitis, fungal urinary tract infection. vulvo-vaginal candidiasis, Tinea pityriasis versicolor, otomycosis.
- *Adverse effects*—Chills, fever, itching, nausea, rashes, phlebitis, anemia, irritation to skin.

Ketoconazole

It was first orally effective azole antifungal agent used in the treatment of systemic fungal infection. It is less toxic than Amphotericin B.

- *Trade name*—Nizoral, Tocon, Fungicide, Funazole.
- *Preparation available*—Oral: Tab 200 mg, Topical: 2% cream, and shampoo.
- *Mechanism of action*—same as Cotrimazole.
- *Indication*—
 - *Mucocutaneous candidiasis*—Tab 200 mg once a day is very effective.
 - *Paracoccidioides mycosis and Chromoblastomycosis*—for adults, 200-400 mg once daily preferably after food; for children after 2 years, 3.3 to 6.6 mg/kg once daily.
 - *Histoplasmosis*—because of toxicity of amphotericin B, ketoconazole is mostly preferred in histoplasmosis.
 - *Leishmaniasis*—oral ketoconazole 600 mg/day for 28 days is most effective in cutaneous Leishmaniasis.
 - *South America blastomycosis / Almeida's disease*—Ketoconazole has been successfully tried in the recent year.
 - *Dermatophytosis, candidiasis, seborrheic dermatitis*—1) Oral dose of 200 mg daily for 2-3 weeks, can be given. 2) Ketoconazole (Nizoral) available as a cream for topical treatment. 3) Shampoo 1% is used for seborrheic dermatitis (Nizoral AD Shampoo).
 - *Ocular infection*—ketoconazole 200 mg tab containing ointment are used.
- *Adverse effect*—nausea, vomiting, loss of appetite, giddiness, headache, rashes, photophobia, paresthesia. It interferes with biosynthesis of adrenal and gonadal steroid hormone producing gynaecomastia, infertility, menstrual irregularities and GIT upset.

Fluconazole

It is new antifungal drug which is more water soluble and has good penetration into cerebrospinal fluid, ocular fluid, vaginal tissue and saliva. It can be administered intravenously or orally.

- *Trade name*—SYSCAN, DIFLUCAN.
- *Preparation*—Oral 50, 100, 150, 200 mg tablets, Topical: powder for 10, 40 mg/ml suspension. Parenteral-2 mg/ml in 100 and 200 ml vial.
- *Mechanism of action*—it acts by inhibiting the enzyme demethylase resulting in the damage to fungus cell membrane. It has fungistatic action.
- *Indication*—
 - *Cryptococcal meningitis*—Fluconazole is drug of choice for treatment and secondary prophylaxis of cryptococcal meningitis. Drug available in oral/

intravenous formulation at the dosage of 100-800 mg/day. For fungal prophylaxis: 50-100 mg once a day.

- *Mucosal candidiasis/mucocutaneous candidiasis*—Ketoconazole can be given orally in the dose of 50-100 mg daily for 14-30 days or intravenously. For mucosal infection, topical application may be useful.
- Systemic candidiasis—for systemic infection, 400 mg of ketoconazole either orally or intravenously on first day then 200-400 mg once a day for 15-30 days. For children: 3-6 mg/kg BW daily in severe life threatening infection in over 1 year.
- Coccidioid disease.
- Prophylactic agent for bone marrow transplantation recipients and AIDS patient. Prophylactic use of fluconazole to reduce fungal infection are used
- *Dermatophytic infection*—alternate day doses of 100-200mg are sufficient to treat them.
- *Sporotrichosis, histoplasmosis, vaginal candidiasis*—200 mg on first day followed by 100 mg daily once a day are quite effective to reduce fungal infection.
- *Adverse effect*—nausea, headache, abdominal pain.

Itraconazole

It is newer antifungal drug which is administered orally. Its efficiency is enhanced in combination with flucytosine.

- *Trade name*—ITROLE, CANDITRAL, SPORANOX.
- *Preparation available*—Oral: 100 mg, 200 mg Capsule. Topical: 10 mg/ml solution. Parenteral: 10 mg/ml for IV infusion.
- *Mechanism of action*—same as Fluconazole.
- *Indication*
 - *Oropharyngeal candidiasis*—oral liquids and intravenous preparation are quite effective in the dose of 200 mg capsule/tablets twice daily for at least 6 month, yields good results.
 - Dermatophytoses treatment
 - *Onychomycosis*—dose of 200 mg daily taken with the food to ensure maximum absorption for 3 consecutive months.
 - *Other*—Aspergillosis, histoplasmosis, sporotrichosis and blastomycosis.
- *Adverse effect*—peripheral neuropathy, headache, dizziness, allergic reaction, jaundice, hepatotoxicity.

Terbinafine

It is new systemic antifungal agent. It has higher efficiency comparable to itraconazole with decreased risk of hepatotoxicity.

- *Trade name*—LAMISIL.
- *Preparation available*—Topical: 1% cream. Oral: 10 mg (tab 250 mg/ day orally).

- *Mechanism of the action*—Terbinafine mainly act as fungicidal. It binds with ergosterol in the cell membrane of fungi, increases the permeability of cell membrane and allows the leakage of macromolecules and anions from cell results in fungal cell death.
- *Indication*—
 - Onychomycosis of the fingernails (Oral preparation in the dose of 250 mg once a day for 6 weeks), onychomycosis of toe nails (oral preparation—250 mg once a day for 12 weeks. 1% cream is used for local application once or twice a day).
 - Dermatophytic infection (topical application of 1% cream for 1 week, but should not exceed 4 weeks).
- *Adverse effect*—local irritation, stinging, dryness.

Newer Antifungal Drugs

- *Silver sulfadiazine (SSZ)*—SSZ has broad spectrum of antifungal activity against Candida, Aspergillus, Fusarium, other fungal infections of the eye.
 - *Trade name*—APLICAPS.
 - *Preparation*—Topical: 1% suspension, Oral: 250 mg tablets.
 - *Indication*—it is used in fungal infection of the eye caused by Candida, Aspergillus, and Fusarium. 1% suspension is used, 5 times a day. SSZ is also available in 1% eyedrops and ointment 1%—applied topically 4-5 times daily.
- *Tolnaftate*—it is newer antifungal drug used topically in the form of creams, powder. It is active against only growing cells. It is resistant to Candida, i.e. *Candida albicans*.
 - *Preparation*—Topical: solution, cream, gel, aerosol powder.
 - *Indication*—it is used in dermatomycophytosis—Topically Gel, Solution or aerosol powder mixed with glycerol is applied over affected area of skin twice a day). It is also used in tinea cruris and tinea corporis. Topical application may be useful.
 - *Adverse effect*—allergic reaction, irritation.
- *Naftifine*—it is newer antifungal drug highly active against dermatophytes.
 - *Preparation*—it is available in 1% cream, gel. Contact with mucous membrane should be avoided.
 - *Indication*—Dermatomyophytosis—Topically applied over the skin is quite effective.
- *Cyclopirox olamine*—it is newer antifungal agent with high cure rates. It has good penetration power.
 - *Trade name*—LAPROX.
 - *Preparation*—1% cream, lotion.
 - *Indication*—it is used in Tinea infections, Dermatophytosis, Candidiasis—Topical application of cream or lotion over skin and oral mucosa, 4-5 times

a day yield good results due to good penetration power.

- *Benzoic acid*—Application of this must be continued till infected keratin is shed. It is used in hyperkeratotic lesion—“*Whitfield’s* ointment contains benzoic acid 5%, salicylic acid 3%. It is used in dermatological infection caused by fungus and bacteria.
- *Undecylenic acid*—it is newer fungistatic agent used in combination with zinc salts.
 - Trade name—CRUEX, DENSENEX.
 - Preparation—10-25% powder, cream, ointment.
 - Indication—It is used in tinea cruris, tinea pedis, skin rash and superficial mycoses.
- *Haloprogin*—it is newest drug having antifungal activity.
 - Trade name—HALOTEX.
 - Preparation—1% cream, solution.
 - Indication—Ringworm infection, Dermal candidiasis.
- *Voriconazole*—it is newest trizole to enter clinical trials. It is available in an intravenous and oral formulation. Recommended dosage is 400 mg/day. It has broad spectrum of action mainly against *Candida* species.

Voriconazole is most effective than itraconazole in candidial infection.

- *Indication*—Oral thrush: Orally or intravenously administered in the dose of 400 mg/day. *Mucocutaneous reparation*—Topical 10% ointment, cream.
- *Indication*—tinea corporis, superficial mycosis and hyperkeratinised lesion: used in combination with salicylic acid.
- *Sodium thiosulfate*—it is weak fungistatic agent useful in pityriasis versicolor. It is not effective in superficial mycosis.
 - Trade name—KARPIN
 - Preparation—2% lotion, solution.
 - candidiasis, dermatophytosis.
- *Butenafine*—it is newer antifungal agent used mostly in the treatment of superficial dermatophytosis.
 - Trade name—MENTAX.
 - Preparation—Topical 1% cream or lotion.
- *Buclosamide*—It is less effective than imidazole.
 - Trade name—JADIT.

Summary Chart

Drug	Trade name	Dosage form	Dose	Indication	Contraindication	Adverse effect
Amphotericin B	Fungizone	Tablet Injection	50-100 mg QID 0.3 mg /kg body weight 4-8 hourly as an infusion	Oral, vaginal cutaneous candidiasis, otomycosis, systemic mycosis, intestinal moniliasis, kala azar	Renal impairment, Epilepsy	Acute reaction, chills, fever, nausea, vomiting, long term nephrotoxicity
Nystatin	Mycostatin	Tablet	5-10 lac units 2-3 times daily	Moniliasis, kala azar, oral candidiasis, intestinal moniliasis	Hypersensitivity	Nausea, diarrhea and bad taste
Hamycin	Hamycin	Suspension	2 lac units /ml 2-3 times daily	Oral thrush	Hypersensitivity	Sensitization, irritation
Griseofulvin	Dermofulvin Fungal 500	Tablet	500-1000 mg daily IM	Fungal infection of skin, nail and scalp	Porphyria, severe liver disease, systemic lupus erythematosus	Headache, irritation
Ketoconazole	Nizral Tocon	Tablet	200 mg OD/BD	Systemic candidiasis, Dermatormycosis	Children below 2 yrs	Hepatitis, rashes, GIT upset
Terbinafin	Exifine Sebifin	Tablet	250 mg /day OD 2-6 weeks	Fungal infections of skin, nails and onychomycosis	Hypersensitivity, Liver dysfunction	GIT symptoms, skin rashes and reaction
Fluconazole	F-zole Fluzone	Capsule Tablet Infusion	200 mg on 1st day followed by 100 mg OD	Systemic vaginal and mucosal candidiasis	Hypersensitivity	Nausea, headache, skin rashes and abdominal pain
Itraconazole	Itrole Canditral	Capsule	100-200 mg OD for 6 months	Oropharyngeal candidiasis, Systemic mycosis, Chronic mucocutaneous endocrinopathy	Hypersensitivity	Nausea, headache, skin rashes and abdominal pain

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Anticancer Drugs

5

Introduction

Anticancer drug is used in the treatment of malignant tumors. It selectively kills tumor cells by virtue of cell kinetic proliferation character and cell biology. It produces only temporary or partial tumor regression. It may be used in combination with radiotherapy or surgery or as palliative treatment where the treatment of cancer is not possible.

Factors Affecting Chemotherapy

- *Cells synthesizing DNA*—response to chemotherapy is proportional to the number of cells synthesizing DNA.
- *Doubling time*—the shorter the doubling time at the onset of treatment, the better is the response as more cells will synthesize DNA.
- *Size of tumor*—as tumor grows, it becomes difficult to cure.

Kinetic Classification of Anticancer Drugs

- *Phase dependent*—it kills cells exponentially at a lower dose but reaches the plateau (a state of little or no changes following a period of activity or progress) at a higher dose. As they can kill cells only during a specific part of the cell cycle they are also called as 'cell cycle specific drugs', e.g. G₁ (vinblastine), S (hydroxy urea), G (daunorubicin), and M (vinblastine).
- *Non-phase dependent*—it kills cells exponentially with an increasing dose and is especially toxic for both; cells in all stages of the cycle and those in G₀ phase, e.g. nitrogen mustard, cyclophosphamide, chlorambucil, dacarbazine.

Principles of Chemotherapy

The Cells Kill Hypothesis of Skipper

Skipper formulated some principles while studying tumor cells in mice suffering from leukemia which showed that:

- The survival of an animal was inversely related to the tumor burden.
- A single leukemic cell is capable of multiplying and killing the host.
- For most drugs, there is a clear relationship between dose of the drugs and eradication of tumor cells.
- A given dose of a drug kills a constant fraction of cells and not a constant number. The same amount of drug is required to reduce the burden from one million cells to 10 cells as from 1 lacks to 1 cell.
- Implication of this cell kill hypothesis is that tumors are best treated when they are small in volume and the treatment must continue until the last cell is eradicated.
- If the treatment is discontinued as soon as the tumor is no longer clinically detectable, at least 10⁹ tumor cells will remain unkilld and relapse is inevitable.

The Norton Simon Hypothesis

- In the tumor, which show Gompertzian type of growth curves, the rate of regrowth increases as the tumor shrinks with the therapy.
- Thus, the level of treatment adequate to initiate a regression may be insufficient to maintain the regression and produce cure. So to overcome this, Norton and Simon hypothesized that to counteract the slowing rate of regression in a tumor responding to therapy, it is necessary to increase the intensity of treatment as the tumor becomes smaller.
- Some time radiotherapy and marrow transplantation can be used to intensify the treatment.

- Multidrug regimens are used to attack residual population of cells, biochemically resistant to the initial combination of drugs.

The Goldie Goldman Model

In 1979 Goldie and Goldman produced a model to explain the development of resistance to anticancer drug by the tumor cells and suggested that the population of cells within the tumor were capable of randomly becoming resistant to the cytotoxic agents by means of spontaneous mutation.

Use of Anticancer Drugs

- *Cure of prolonged remission*—this is now possible in acute leukemia, Wilm's tumor, Ewing sarcoma, retinoblastoma, rhabdomyosarcoma, choriocarcinoma, lymphosarcoma, Burkitt's lymphoma and testicular teratomas.
- *Palliative treatment*—it gratify results by shrinkage of tumors, alleviation of symptoms. Life is prolonged in breast cancer, prostatic carcinoma, and small cell cancer of lung, chronic lymphatic leukemia, chronic myeloid leukemia and non-Hodgkin's lymphoma.
- *Adjuvant chemotherapy*—these are used to mop up any residual malignant cells after surgery or radiotherapy.
- *Combination chemotherapy*—anticancer drugs can be used in combination. The drug should be active against the tumor when use alone, should have different mechanism of action and have minimally overlapping toxicity. By using several drugs which attack the cells in different ways it is hoped that the development of resistant cells will be inhibited.

Classification

Alkylating Agents

- Alkyl Sulfonate—Busulfan
- Nitrogen Mustards—Mustine HCl, Cyclophosphamide, Melphalan and Chlorambucil
- Nitrosoureas—Carmustine (BCNU), Lomustine (CCNU), Semustine (Methyl-CCNU)
- Triazine—Dacarbazine
- Ethylenimine—Thio- TEPA

Anti-Metabolites

- Purine antagonist—6-Mercaptopurine, 6-Thioguanine
- Folate antagonist—Methotrexate
- Pyrimidine antagonist—5-Fluorouracil, Cytarabine (cytosine arabinoside).

Antibiotics

- Actinomycin-D

- Mitoxantrone
- Bleomycin
- Daunorubicin
- Doxorubicin
- Mithramycin
- Mitomycin C

Vinca alkaloids

- Vinblastine
- Vincristine

Taxanes

- Paclitaxel
- Docetaxel

Epipodophyllotoxin

- Etoposide
- Teniposide

Enzymes

- L-Asparaginase

Miscellaneous

- Hydroxyurea
- Procarbazine
- Cisplatin
- Hexamethylamine
- Carboplatin

Alkylating Agents

They produce their effect by linking an alkyl group ($R-CH_2$) covalently in protein and nucleic acid. Some of them are described below:

Alkyl Sulfonate

- *Busulfan*—It is highly specific for myeloid elements, granulocyte precursors being most sensitive, followed by those of platelets and RBCs.
- *Use*—It is the drug of choice of chronic myeloid leukemia.
- *Dose*—2-6 mg/day orally.

Nitrogen Mustards

- *Mechanism*—it has two chloroethyl side chains. One of them forms a cyclical highly reactive ammonium ion which binds to nucleic acids, i.e. 7-nitrogen group of guanine. The other chloroethyl sides of nitrogen mustards can crosslink with DNA strands, either within a strand or between strands. Although alkylating agents may bind to variety of cellular components like cytoplasmic proteins and RNA at therapeutic doses, impairment of DNA replication is the major mechanism of cytotoxicity of these drugs. Damage to DNA is more serious during the 'S' phase of cell cycle probably

because the cell has less time to excise the damage to DNA fragment.

- *Mustine HCl*—It is the first nitrogen mustard, highly reactive and local vesicant. Dose—0.4 mg/kg IV in 1-4 days.
- *Cyclophosphamide*—it has prominent immunosuppressant property. Dose—2-3 mg/kg/day orally, 10-15 mg/kg IV every 7-10 days.
- *Chlorambucil*—it is the slow acting alkylating agent, especially active on lymphoid tissue. It is drug of choice for chronic lymphatic leukemia. Dose—4-10 mg daily for 3-6 weeks, then 2 mg daily for maintenance.
- *Melphalan*—it is very effective in multiple myeloma. Dose—10 mg daily for 7 days or 6 mg/day for 2-3 weeks and after 4 weeks' gap, 2-4 mg daily for maintenance orally.

Nitrosoureas

- *Mechanism*—they act partly as alkylating agents linking to an alkyl group or carbonyl group of cell proteins and form compounds which are unstable in water and decompose to form alkylating groups, which are able to damage the cell proteins.
- *Dose*—BCNU—50-200 mg/m², CCNU 100-130 mg/m², methyl CCNU—100-200 mg/m².

Triazine

- *Dacarbazine*—it is different from other alkylating agents in having primary inhibitory action on RNA and protein synthesis. It is activated in the liver. It is used in malignant melanoma. Dose—3.5 mg/kg/day IV for 10 days, repeat after 4 weeks.

Ethylenimine

- *Thio-TEPA*—it does not require formation of an active intermediate. It has high toxicity and seldom used today. Dose—0.3-0.4 mg/kg IV at 1-4 weeks intervals.

Anti-metabolites

Purine Antagonist

- *Mechanism of action*—Mercaptopurine and thioguanine are highly effective anti-cancer drugs. It inhibits conversion of inosine monophosphate to adenine and guanine nucleotides.
- *Use*—they are useful in childhood acute leukemia and choriocarcinoma.
- *Toxic effect*—main toxic effects of antipurines are bone marrow depression which develops slowly.
- *Dose*—6-Mercaptopurine (2.5 mg/kg/day) and 6-Thioguanine (2 mg/kg/day).

Folate Acid Antagonist

- *Action*—Most commonly used folic acid antagonist is methotrexate. It inhibits the conversion of dihydrofolate to tetrahydrofolate which in turn is converted to a variety of coenzymes. It blocks thymidylate monophosphate synthesis and thus inhibiting RNA and DNA synthesis and so methotrexate is 'S' phase specific.
- *Absorption*—it is well absorbed from the gut at lower doses (up to 100 mg) but higher doses should be given intravenously. After IV injection, there is a rapid early half life of 45 minutes, a slower phase of renal excretion of about 3 hours and then a very long terminal half. Methotrexate does not penetrate the CSF at conventional doses. To ensure adequate levels in CSF, methotrexate may be given intrathecally at a dose of 10 mg/m².
- *Toxicity*—the prolonged clearance of methotrexate is responsible for the toxicity to marrow gut and mucous membrane which leads to bone marrow depression, diarrhea and oral ulceration. The toxicity of methotrexate can be reversed by folic acid.
- *Contraindications*—it should be avoided in patients with ascites or pleural effusion as the drug may accumulate in fluid reservoirs and will release slowly causing toxicity.
- *Indications*—Used in treatment of acute leukemia, non-Hodgkin's lymphoma, breast cancer and osteogenic sarcoma.
- *Dose*—in choriocarcinoma 15-30 mg/day for 5 days orally or 20-40 mg/m² IM or IV twice a week. In maintaining remission in children with acute leukemias, 2.5-15 mg/day is useful.

Pyrimidine Antagonists

- *Action*—most commonly used are 5-fluorouracil and cytarabine. It interferes with nucleic acid synthesis by antagonizing or mimicking pyrimidine metabolites.
- *Absorption*—it may be given orally but its absorption is unpredictable. So, the IV route is often used because the plasma clearance is rapid.
- *Toxicity*—nausea, vomiting, stomatitis, alopecia and myelosuppression.
- *Indications*—it is given in the treatment of breast and GIT cancer, Hodgkin's lymphoma, non-Hodgkin's lymphoma, acute leukemia to induce remission.
- *Dose*—fluorouracil—1 gm orally on alternate day for 6 days followed by 1 gm weekly or 12 mg/kg/day IV for 4 days. Cytarabine—1.5-3 mg/kg IV BD for 5-10 days.

Antibiotics

Practically all of the antibiotics intercalate between DNA strands and interfere with template function.

Actinomycin-D

- **Action**—it is isolated from streptomyces. It is intercalated between guanine and cytosine base pairs and blocks the transcription of DNA and DNA related RNA synthesis. It inhibits division of rapidly dividing cells. It is given intravenously and plasma clearance is within a few minutes.
- **Toxicity**—nausea, vomiting, mucositis, diarrhea and myelosuppression.
- **Indications**—it is active against rhabdomyosarcoma, Wilm's tumor Ewing's sarcoma, and teratoma.
- **Dose**—it may be given as a single injection of 15 µg/kg IV daily for 5 days or in combination with cyclophosphamide and vincristine.

Mitoxantrone

- **Action**—it binds to DNA and is given intravenously and has a long terminal half life of 36 hours.
- **Toxicity**—it may cause myelosuppression, cardiomyopathy and alopecia.
- **Indications**—acute non-hemolytic leukemia, chronic myelogenous leukemia, non-Hodgkin lymphoma and carcinoma of breast.
- **Dose**—12 mg/m² single IV dose repeat at 3 weeks.

Bleomycin

- **Action**—It consists of a mixture of closely related glycopeptide antibiotics. It inhibits DNA synthesis and causes a break in DNA and is active in G₂ phase of the cell cycle.
- **Absorption**—It is given by parenteral, subcutaneous or intramuscular route because it is non-vesicant. It has initial half life of 30 minutes and elimination from plasma takes 2-9 hours. It is excreted by kidney and does not cross the CSF.
- **Toxicity**—skin pigmentation, erythema, vesiculation or fibrosis.
- **Indications**—it is used in combination with other chemotherapeutic drugs in testicular carcinoma, head and neck cancer and Hodgkin's lymphoma.
- **Dose**—30 mg per injection twice weekly IV.

Doxorubicin and Daunorubicin

- **Action**—These are antitumor antibiotics and are produced by streptomyces fungus. They are capable of causing breaks in DNA strands by activating topoisomerase II and generating quinone type radicals.
- **Absorption**—they are given by IV route and cleared from plasma, metabolized in liver and excreted in bile. So great care should be taken while prescribing in liver dysfunction patients.

- **Toxicity**—nausea, vomiting, alopecia and diarrhea occur. Cardiotoxicity can occur which manifests as ECG changes, arrhythmias and hypotension.
- **Indications**—doxorubicin is given in lymphoma (Hodgkin's disease), small cell cancer, breast cancer and daunorubicin is given in acute myeloid and lymphatic leukemia. Daunorubicin used is limited to acute leukemia.
- **Dose**—doxorubicin—60-75 mg/m² IV every 3 weeks and daunorubicin—30-60 mg/m² IV daily for 3 days and repeat weekly.

Mithramycin

- **Action**—its use is restricted to embryonal testicular tumor, disseminated cancers, especially those with bony metastasis and hypercalcemia. It reduces serum calcium levels, probably by direct action on bone inhibiting calcium release.
- **Dose**—25 µg/kg by slow IV infusion daily or on alternate days.

Mitomycin-C

- **Action**—it is derived from streptomyces species and inhibits DNA synthesis by both cross linking and alkylating DNA.
- **Toxicity**—myelosuppression, cumulative thrombocytopenia and renal toxicity.
- **Indications**—used in combination in cancer of breast, stomach, cervix, pancreas and those of head and neck.
- **Dose**—10 mg/m² IV.

The Vinca Alkaloids

They are mitotic inhibitors, which bind to 'tubulin' (proteins of the cellular microtubules) to cause disruption of mitotic spindle and interfere with cytoskeletal function. The vinca alkaloid blocks assembly of A and B subunit of the tubulin preventing the formation of the microtubules.

Vinblastine

- **Absorption**—these drugs are given IV and are vesicants if extravasated. Plasma clearance occurs in 3 phases with half life of 4 minutes, 1 hour and 16 hours. Tissue binding is extensive and prolongs the actions and the drug binds to platelets, red cells and plasma proteins.
- **Indications**—it is used in combination for treatment of testicular tumors and lymphoma.
- **Toxicity**—it causes alopecia, neurotoxicity and myelosuppression.
- **Dose**—0.1-0.15 mg/kg IV weekly in 3 doses.

Vincristine

- *Use*—it is a rapidly acting drug, very useful for inducing remission in childhood acute leukemia.
- *Toxicity*—it causes peripheral neuropathy, alopecia.
- *Indications*—it is medicated in lymphoma (Hodgkin's disease), acute lymphatic leukemia, small cell cancer of bronchi and breast cancer.
- *Dose*—1.5 mg/m² IV weekly.

Taxanes**Paclitaxel**

- *Action*—it is a complex diterpin taxane from bark of the Western yew tree. It enhances polymerization of tubulin. The microtubules are stabilized and their depolymerization is prevented.
- *Indications*—it is used in metastatic ovarian and breast carcinoma after failure of first line chemotherapy and in relapse cases. It can be used in head and neck cancer, small cell lung cancer.
- *Dose*—175 mg/m² by IV infusion over 3 to 24 hours repeated every 3 weeks.

Docetaxel

It is a congener of paclitaxel with the same mechanism of action. It has been found to have efficacy in metastatic breast cancer refractory to first line drugs. Major toxicity is neutropenia.

Epipodophyllotoxin

- *Action*—these are phase specific and prevent cells from entering mitosis from G₂ phase.
- *Absorption*—the drugs are absorbed erratically from the gut with the plasma availability up to 50%, rapid clearance from plasma occurs when given intravenously followed by a slower phase.
- *Etoposide*—it is highly protein bound and is excreted in the urine in 72 hours.
- *Teniposide*—it is active as a single agent in small cell lung cancer.
- *Toxicity*—they cause alopecia, myelosuppression, mucositis and neuropathy.
- *Indications*—it is used in treatment of testicular tumors, leukemia and lymphoma.
- *Dose*—100 mg in 5 ml injection or 120 mg/m² IV infusion for 30 minutes.

Enzymes

- *L-Asparaginase*—this enzyme is produced by *E. coli*.
- *Action*—it acts by removing asparagine from the circulation, thus depressing those tumor cells which are

unable to synthesize asparagine due to lack of or have very low levels of asparagine synthetase.

- *Route of administration*—drug is usually given intravenously after a skin test for hypersensitivity.
- *Indications*—it is used in the remission period in acute lymphocytic leukemia.
- *Toxicity*—anaphylaxis, pancreatitis, hypoglycemia, hypoproteinemia, encephalopathy and nausea.
- *Dose*—50-200 KU/kg IV daily for 2-4 weeks.

Miscellaneous Agents**Hydroxyurea**

- *Action*—it blocks the conversion of ribonucleotides to deoxyribonucleotides by inhibiting the enzyme ribonucleoside diphosphate reductase interfering with DNA synthesis. It is 'S' phase specific. It is well absorbed orally and crosses the CSF.
- *Toxicity*—it causes neutropenia and gut disturbance. There is also myelosuppression.
- *Indications*—it is used in chronic granulocytic leukemia, polycythemia vera.
- *Dose*—20-30 mg/daily for 8 weeks.

Procarbazine

- *Action*—this is a weak mono-aminoacid oxidase inhibitor which inhibits action of DNA and RNA and depresses proline synthesis. It also causes chromosomal damage.
- *Absorption*—it is well absorbed from the gut and is one of the few drugs which penetrate the CSF.
- *Indications*—it is used mainly in Hodgkin's disease and brain tumor.
- *Toxicity*—it includes nausea, vomiting and leukopenia.
- *Dose*: 100 -300 mg orally daily for 2 weeks.

Cisplatin

- *Action*—this drug is the only active cytotoxic agent in its cis form. Chloride ions are lost from the molecule after it diffuses into the cell and the compound crosslinks mainly to guanine groups like an alkylating agent. It should be protected from light and is given intravenously with an early half life of about 40 minutes with a later slower phase of clearance i.e. about 60 hours.
- *Absorption*—about 90% of cisplatin bound to plasma protein is taken up in the kidney, gut, liver, testis and ovary, but it does not cross the blood-brain barrier. It is excreted by the kidney.
- *Toxicity*—it is nephrotoxic, ototoxic and may cause severe nausea and vomiting. Renal damage may be cumulative. Magnesium wasting may occur as a result of renal damage. If it is given in large doses it is associated with peripheral neuropathy, predominately affecting sensory nerve endings.

- **Indications**—it is very effective in testicular tumors and in ovarian cancer. It is also effective in bladder, head and neck tumors, small cell cancer of bronchi and osteosarcoma.
- **Dose**—50-100 mg/m² every 3-4 weeks.

Hexamethylamine

- **Indications**—it is active against ovarian and cervical cancer.
- **Absorption**—it is well absorbed from the gut.
- **Toxicity**—it causes abdominal cramps, diarrhea and leucopenia. CNS toxicity includes altered mental state and convulsions.
- **Dose**—it is given orally at a dose of 12 mg/kg/day for 14 days.

Carboplatin

- **Action**—it is a less reactive second generation platinum compound that is better tolerated.
- **Toxicity**—nephrotoxicity, ototoxicity and neurotoxicity are low. The dose limiting toxicity is thrombocytopenia and less often neutropenia.
- **Indications**—it is primarily indicated in ovarian carcinoma of epithelial origin, squamous cell carcinoma of head and neck, small lung cancer and *seminoma*.
- **Dose**—400 mg/m² as an IV infusion over 15-60 minutes to be repeated only after 4 weeks.

Pharmacology

The susceptibility of a tumor cell to drugs depends on the sensitivity of the tumor cells to action of the drug, whether the cell is in cycle or not and whether the drug reaches the cell at high enough concentration for a sufficient time to effect cell kill. The concentration of cytotoxic drugs at the tumor site, duration and integrity of its action are determined by several factors such as drug absorption, binding, distribution, metabolism, excretion tumor size and its vascularity. The higher dose is better in terms of response and relapse free interval and overall survival.

Principles of Treatment

Routes of Administration

The route of administration of a cytotoxic drug is determined by the stability, size, molecular charge and sclerosant characteristics of that drug. The intravenous route is most commonly used because a known concentration of the drug is delivered directly to the central component of the tumor.

Oral route is used infrequently because of the unpredictability of patient compliance and variable absorption of the drug. Subcutaneous and intramuscular route are rarely employed because of drug instability and the risk of thrombocytopenia and because large volumes of diluent therapy may be required to dissolve the drug. Intraperitoneal injection is beneficial for the drug delivered to small tumor nodules on the peritoneal surface, but maintenance of access is the major problem with this approach.

Combination Chemotherapy

Three general principles govern the use of combination chemotherapy. The drugs should be:

- Active against the tumor when used alone.
- Have different mechanism of action.
- Have minimally overlapping toxicities.

By using several drugs which attack the cells in different ways it is hoped that the development of resistant cells will be inhibited. If the above three guidelines are used then the dose chosen for each individual drug will be closed to that used when the drug is given as a single agent resulting in a minimum reduction in dosage intensity.

Schedule

The majority of intravenous drugs are given as intravenous injections or as a short infusion. Longer infusions may be preferable either to reduce the incidence of toxicity or to increase the efficiency.

Multimodality Treatment and Timing of Chemotherapy

At earlier stages of the tumor, the growth fraction is higher, spontaneous resistance may be less, tumor load is smaller and the drug penetrates a smaller volume greater, so chemotherapy is effective and preferred in early tumors. Before starting the treatment, prognostic factors for tumor type should be studied and the adjunct chemotherapy is selected. The chemotherapy given before the definite primary treatment is called as neoadjuvant chemotherapy. Its aim is to reduce tumor bulk to permit more radical local therapy. Radiotherapy may be combined with chemotherapy.

Toxicity

Many cytotoxic drugs are associated with side effects at commonly used therapeutic doses. Long term side effect of chemotherapy result in an increased risk of secondary malignancy and infertility in some cases.

Local Toxicity

Some cytotoxic drugs cause severe local reaction when extravasated, e.g. daunorubicin, mitomycin and mustine HCl. The use of trained personnel for injection of cytotoxic drugs reduces the hazards of administration.

Hematological Toxicity

Bone marrow suppression is the most important dose limiting toxicity. Myelosuppression is expected to be maximum in 10-14 days after treatment. Certain drugs such as mitomycin and nitrosoureas have a delayed nadir at 4-5 weeks. Hence, these drugs cannot be given more than once in week. Some of the alkylating agents have a cumulative effect on the bone marrow stem cells, e.g. chlorambucil, busulphan and melphalan. So the count may fall gradually for several weeks and recovery is slow.

Gastrointestinal Toxicity

The precise cause of nausea and vomiting which is commonly seen with cytotoxic drugs is uncertain; but it is probably due to a combination of stimuli from the chemoreceptor trigger zone. The timing of onset of vomiting varies and may occur within 2 hours. Prophylactic antiemetics are used to abolish vomiting and reduce nausea. Vinca alkaloids may cause constipation and paralytic ileus which will usually resolve spontaneously.

Alopecia

Generally, the head hair is lost but the whole of the body hair may be affected. The hair follicles are affected because of the high rate of cell turnover. Hair loss due to daunorubicin may be reduced by scalp cooling which by causing local vasospasm reduces the amount of drug reaching the follicles.

Pulmonary Toxicity

It is associated with only a few cytotoxic agents such as bleomycin, busulphan, cyclophosphamide and methotrexate. The pulmonary changes with infiltrate may be transient or may progress to pulmonary fibrosis.

Cardiac Toxicity

Cardiomyopathy may be seen with certain drugs. The exact cause is uncertain. Cardiac arrhythmias may be seen during or recently after the injection of daunorubicin.

Renal and Bladder Toxicity

Cisplatin may cause nephrotoxicity leading to fall in glomerular filtration rate and tubular dysfunction with subsequent hypocalcemia and hypomagnesemia. High doses of methotrexate may cause renal damage. The damage may be avoided by treating only those patients with satisfactory renal function.

Neurological Toxicity

The most common toxicity associated with cytotoxic drugs is peripheral neuropathy as seen with vinca alkaloids. Loss of tendon reflexes, paresthesia and numbness in the finger and toes may be noted only and are an indication to reduce the dosage. Development of myalgia, neuritic pain and peripheral sensory loss is an indication to stop the treatment. Drowsiness, confusion and encephalopathy may be seen with cyclophosphamide, procarbazine and *dacarbazine*.

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Introduction

Viruses are the ultimate expression of parasitism. They take nutrition from the host cell and also direct its metabolic machinery to synthesize new virus particles. It consists of double stranded or single stranded DNA or RNA enclosed in protein coat called capsid.

Some viruses contain enzymes that initiate viral replication in the host cell. The whole infective particle is a virion. Their replication depends primarily on synthetic processes of the host cell. They not only take nutrition from the host cell but also direct its metabolic machinery to synthesize new viral particles.

Antiviral agents are most active when viruses are replicating. Viral replication consists of several steps like

- Adsorption to and penetration to susceptible host cells.
- Uncoating of viral nucleic acid.
- Synthesis of early regulatory protein/structural protein.
- Synthesis of RNA and DNA.
- Assembly/maturation of viral particles.

Drugs can potentially target any these steps, so antiviral drugs have to be specific for the different viral mechanism. In many viral infections, replication of virus peaks at or before the manifestation of clinical symptoms. To be effective, therefore, therapy has to be started in the incubation period, i.e. has to be prophylactic.

Action of Antiviral Drugs

Antiviral agents must either block viral entry into or exit from the cell or be active inside the host cell. Antiviral agents must inhibit virus specific directed nuclei rather than host cell directed nucleic acid. As nonselective inhibitors of virus replication may interfere with host cell function and produce toxicity, the search for chemicals that inhibit virus-

specific function is currently one of the most active areas of pharmacological investigation. Optimal efficacy, therefore, generally depends either on early initiation of therapy or on prevention of infection.

Classification

According to their therapeutic use

- *Anti-herpes virus agents*
 - Acyclovir
 - Idoxuridine
 - Gancyclovir
 - Trifluridine
 - Vidarabine
 - Pencyclovir
 - Famcyclovir
 - Valcyclovir
 - Foscarnet
- *Anti-retroviral agents*
 - Reverse transcriptase inhibitors
 - Zidovudine (AZT)
 - Didanosine
 - Zalcitabine
 - Stavudine
 - Protease inhibitors
 - Saquinavir
 - Indinavir
 - Nelfinavir
 - Ritonavir
 - Amprenavir
- *Anti-influenza virus agents*
 - Amantadine
 - Rimantadine
- *Nonselective antiviral agents*
 - Ribavirin
 - Lamivudine
 - Human interferon

Antiherpes Virus Agents

Acyclovir

Acyclovir is a synthetic purine nucleoside analogue. It is active against herpes group of virus (HSV-1 and HSV-2). Herpes simplex is the most sensitive to acyclovir followed by varicella zoster. Activity against Epstein-Barr virus, cytomegalovirus and human herpes virus-6 is comparatively weaker.

- **Mechanism of action**

- Antiviral drugs require virus specific enzymes for conversion to active metabolites that inhibits DNA synthesis and viral replication.
- Acyclovir in presence of herpes virus specific thymidine kinase is converted to acyclovir monophosphate which in presence of cellular kinase is converted to acyclovir triphosphate. This acyclovir triphosphate gets incorporated in viral DNA and stops lengthening of DNA strand and inhibits herpes virus DNA polymerase competitively.
- It is preferably taken up by the virus infected cells. Due to selective generation of active inhibitor in the virus infected cells and its greater inhibitory effect on viral DNA synthesis, Acyclovir has low toxicity for host cells.

- **Indication**

- *Herpes simplex mucocutaneous infection*—mucocutaneous H. simplex is a type 1 viral disease remains localized to lips and gums may be treated with acyclovir cream at least for 10 days. Oral lesions can be treated by applying oral suspension or tablet which dissolves in the oral cavity 4-5 times daily for 10 days. This treatment is most effective when new lesions are forming. Prophylactic oral therapy may prevent sun exposure related recurrences. Sometimes, disease often gets disseminated in immunocompromised individuals and may be treated with oral or intravenous acyclovir (15 mg/kg/day) for 7 days.
- *Ocular keratitis*—it is used as an alternative to idoxuridine drug, may be better for deep infections because of good corneal penetration. Eye ointments should be used 5 times daily till for 3 days after healing of corneal lesion.
- *Encephalitis H simplex*—acyclovir 10 mg/kg 8 hourly IV for 10 days is the drug of choice. Treatment will be effective if it is started early to avoid the mortality and neurological complication.
- *Genital herpes simplex*—genital infection can be treated by topical, oral or parenteral acyclovir depending on stage and severity of disease. In primary disease, 5% ointment is applied locally 6 times a day for 10 days is effective. In severe cases, they should receive oral

therapy (1 g/day in 5 divided doses for 7 days) in combination with local therapy. It gives symptomatic relief and rapid healing of lesion. In recurrent disease, topical therapy is quite ineffective. Most of the cases may be treated parenterally by 5 mg/kg IV infused over 1 hour, repeated 8 hourly for 10 days. Suppressive oral therapy with 200 mg 4 times a day prevents the recurrence as long as given.

- *Herpes zoster*—acyclovir is used in the dose of 5-10 mg/kg 8 hour IV for 7 days. Oral therapy is also beneficial if started early. It affords symptomatic relief and faster healing of lesion. Ointment may be applied on herpetic ulcers but postherpetic neuralgia is not prevented.
- *Chickenpox*—acyclovir (15 mg/kg/day for 7 days) is drug of choice. It reduces fever, eruptions, hastens healing and prevents visceral complication.
- *Shingles in immunocompetent person*—Acyclovir is used in the form of tablets or suspension. It gives best result within 48 hour of appearance of rashes. Severe symptoms may require IV administration.
- **Adverse effect**
 - Local reaction at the site of injection with inflammation, pain, phlebitis.
 - Stinging and burning sensation at the site of application
 - Headache, nausea, malaise.
 - Increase in blood level of urea and creatinine.
 - Renal impairment, acute renal failure.
 - Skin rashes
 - GIT disturbance, sweating, convulsions, coma.
- **Trade name**—CYCLOVIR, ZOVIRAX
- **Preparation**—Oral: 200, 400, 800 mg tablets. Topical: 5% ointment, 200 mg/5 ml suspension. Parenteral: powder to reconstitute for IV injection (250, 500 mg/vial).

Idoxuridine

It acts as a thymidine analogue. It was the first pyrimidine antimetabolite to be used as antiviral drug. It is primarily active against DNA virus.

- **Mechanism of action**—it competes with thymidine, gets incorporated into the DNA of virus. Therefore, it disrupts the DNA synthesis and the faulty DNA is formed which breaks down easily. This DNA directs the synthesis of wrong viral proteins (faulty transcription occurs). Clinical utility is limited to herpes simplex keratitis.
- **Indication**
 - *Ocular infection*—it is mainly used for ocular infections.
 - *Herpes simplex keratoconjunctivitis*—one drop of 0.1% solution is put hourly during daytime and 2 hourly during night. 0.5 % eye ointment may be applied

4 hourly for the period of 3 weeks in acute infection. Corneal ulcers respond well to this treatment and blindness can be prevented.

- *Adverse effect*
 - Systemic toxicity (bone marrow depression) is high after IV injections. So, intravenous infusions are avoided.
 - May develop the resistance readily to idoxuridine.
 - Ocular irritation, edema of eyelids, photophobia.
- *Trade name*—RIDINOX
- *Preparation*—topical: 0.1% eye drops, 0.5% eye ointments.

Gancyclovir

It is about 100 times more active against herpes viruses including cytomegalovirus. It is more active than acyclovir intracellular.

- *Mechanism of action*—gancyclovir requires triphosphorylation for activation prior to inhibiting the viral DNA polymerase. Activated compound competitively inhibit viral DNA polymerase and causes termination of viral elongation and inhibit the synthesis of DNA. As the concentration of active triphosphate is much higher inside CMV infected cell. Ganciclovir is much active against CMV, HSV, VZV, and EBV than that of acyclovir.
- *Indication:*
 - *CMV retinitis with AIDS patient*—IV ganciclovir 5 mg/kg every 12 hours for 2 weeks during induction and then 5mg/kg/day for maintenance therapy is useful. Dual therapy with foscarnet and ganciclovir has been shown to be more effective than either drug administered alone. Recently, ganciclovir intraocular implant which release ganciclovir into the vitreous cavity of eye is administered at the rate of 1.4 ug/hour has been proved for treatment of CMV retinitis. Surgical replacement is required at approximately 5 to 8 months intervals. Implant achieves high and prolonged intraocular levels of ganciclovir shown to delay progression of retinitis to a greater degree than systemic therapy.
 - *CMV colitis and esophagitis*—intravenous ganciclovir is used in AIDS patient.
 - *Before any organ transplantation*—IV administration reduces the incidence of symptomatic CMV disease if administered before organ transplantation.
 - *CMV pneumonitis*—in immunocompromised patient, it is often beneficial.
- *Adverse effect:*
 - Systemic toxicity: bone marrow depression.
 - Rash, fever, vomiting.
 - CNS disturbance: headache, seizures.
 - Neutropenia (20-40 % patient).

- *Trade name*—CYTOVENE, VITRASERT
- *Preparation*—Oral: 250 mg capsules. Parenteral: 500 mg/vial for IV injections. Intraocular implants: 4.5 mg ganciclovir/implant.

Famcyclovir

It is an ester prodrug of guanine nucleoside analogue of pencyclovir. It has good oral bioavailability. It has potency against herpes simplex virus and varicella-zoster virus. It is also inhibitory for hepatitis B virus.

- *Mechanism of action*—like acyclovir, famciclovir needs viral thymidine kinase for generation of active DNA polymerase inhibitor to block DNA synthesis. It is also inhibitory for hepatitis B virus.
- *Indication:*
 - *Alternative to acyclovir*—it is used as an alternative to acyclovir for genital herpes and herpes zoster.
 - *Recurrent orolabial HSV infection*—it is used in the dose of 500 mg twice a daily for 7 days.
 - *Shingles*—shingles which caused by herpes zoster is best treated with famcyclovir tablets in the dose of 500 mg every 8 hours for 7 days.
 - *Genital herpes infection*—orally drug 125 mg twice daily for 5 days is required. For recurrent genital herpes in immunocompetent patient, need 250 mg twice daily for upto 1 year.
- *Adverse effect:*
 - Headache, diarrhea, nausea.
 - Itching, rashes, mental confusion.
- *Trade name*—FAMTREX, FAMVIR.
- *Preparation*—Oral: 125, 250, 500 mg tablets.

Foscarnet

It is an inorganic pyrophosphate analogue; inhibit herpes viral DNA polymerase, RNA polymerase and HIV reverse transcriptase directly without activation of phosphorylation. Foscarnet is active against herpes virus, CMV and HIV at 3 µg concentration.

- *Mechanism of action*—drug act on the pyrophosphate binding sites of enzyme polymerase and reversibly blocks the site in a noncompetitive manner and inhibits cleavage of pyrophosphate from deoxynucleotide triphosphate.
- *Indication*—
 - *CMV retinitis*—in CMV retinitis with AIDS patient, 60 mg/kg every 12 hours for induction followed by 90-120 mg/kg/day for maintenance therapy.
 - *Acyclovir resistant mucocutaneous herpes simplex (type 2)*—40 mg/kg every 8 hours or 40 mg/kg every 12 hours yields good results.
 - *Varicella-zoster infection in AIDS patient*—it decreases HIV viral titer but not used primary for HIV infection.

It decreases HIV infection and produces moderate clinical improvement. Combination of zidovudine with foscarnet has been seen to be beneficial in HIV infected patient.

- *Adverse effect:*
 - Toxicity of foscarnet is high. Damage to kidney may produce renal diabetes like condition, acute renal failure.
 - Anemia, phlebitis, tremor, convulsions.
 - Neurological or constitutional symptoms due to hypocalcemia, alteration in PO_4 , K^+ , Mg^{++} .
- *Trade name*—FOSCAVIR
- *Preparation*—Parenteral: 24 mg/ml for IV injection

Antiretroviral Agents

These are the drugs active against human immunodeficiency virus (HIV) which is a retrovirus. They are mainly useful in prolonging life and postponing complication of AIDS or AIDS related complex but not fully cures the disease.

Reverse Transcriptase Inhibitors

Zidovudine Azidothymidine (AZT)

- *Mechanism of action*—cellular enzyme thymidine kinase phosphorylates zidovudine to a triphosphate. Triphosphate competitively inhibits viral RNA-dependent DNA polymerase (reverse transcriptase). Reverse transcriptase produce double stranded DNA copy. This DNA translocates to the nucleus and is integrated with chromosomal DNA of the host cell, which starts transcribing viral genomic RNA and mRNA. Viral regulatory and structural proteins are produced. Zidovudine itself gets incorporated into growing viral DNA and terminates chain elongation. Zidovudine thus prevents infection of new cells by HIV but, has no effect on virus directed DNA that has already integrated into the host chromosome. Zidovudine is effective against retrovirus only. Resistant to AZT can occur by point mutation which alters reverse transcriptase enzyme.
- *Indication:*
 - *AIDS or AIDS related complex*—the mortality and opportunistic infections are reduced by the use of zidovudine in the dose of 200 mg initially six times daily and thereafter, 500 to 1500 mg daily in 4-5 divided doses. In children over three months, 180 mg/sqm four times daily.
 - *Other AIDS associated diseases* such as thrombocytopenia, psoriasis, lymphocytic interstitial pneumonia, 100 mg cap, 300 mg tab to be taken with plenty of water.

- *Neurological diseases related to AIDS, Kaposi's sarcoma*—zidovudine reduces the manifestation and Kaposi's lesions do not appear.
- *Adverse effect*—anemia / granulocytopenia, CNS toxicity is mainly due to partial inhibition of cellular DNA polymerase. Nausea, anorexia, abdominal pain, headache, insomnia, myalgia at initial doses.
- *Trade name*—RETROVIR, ZIDOVIR
- *Preparation*—Oral: 100 mg capsules, 300 mg tablets, 50 mg/5 ml syrup. Parenteral: 10 mg/ml.

Didanosine and Zalcitabine

They belong to the group of dideoxynucleoside class of compounds.

- *Mechanism of action*—they act similar to zidovudine, that is, they inhibit reverse transcriptase after undergoing phosphorylation to generate triphosphate derivatives.
- *Indication:*
 - It is a good alternative to patients resistant to zidovudine.
 - Anti HIV efficacy of these drugs are comparable to zidovudine and hence, it is used in combination regime.
- *Adverse effect*—the most prominent dose related toxicity is pancreatitis and peripheral neuropathy. Diarrhea, abdominal pain, nausea.
- *Trade name*—Didanosine-DINEX, VIDEX. Zalcitabine-HIVID.
- *Preparation*—for didanosine—Oral: 25, 50, 100, 250 mg tab; 100, 250, 375 mg powder for solution; 2, 4 g powder for pediatric solution, for zalcitabine—Oral: 0.75 mg tab.

Protease Inhibitors

As these agent acts at the late stage of HIV replication, they are effective in both newly and chronically infected cells and thus, prevent the infection.

Ritonavir, Indinavir, Nelfinavir

These are specific inhibitor of HIV 1 and HIV 2. Protease must be consumed on an empty stomach for maximal absorption with good oral bioavailability. These drugs are protein bound.

- *Mechanism of action*—as protease enzymes encoded by HIV is involved in the production of structural protein and enzymes (including reverse transcriptase) of virus. These large viral polyprotein is broken into various functional component by this enzymes protease which acts at late step of HIV growing cycle, i.e. maturation of new virus particle. Protease inhibitor drugs bind to protease molecule and interfere with its cleavage function and produce immature noninfectious viral progeny. Hence prevents further round of infections.

- **Indication**—It is mainly used in HIV patients by following dose therapy:
 - Monotherapy with one of the drug in patient treated with AZT reduces HIV viral titre, increased CD4 cell count and improved clinical condition.
 - In PI resistant patient over month, combination of saquinavir with AZT or zalcitabine is more effective than double therapy.
 - Recently, PI in combination with two reverse transcriptase inhibitor (1 nucleotide +1 non-nucleotide) is recommended.
 - In booster dose, PI regimen containing both indinavir and low dose of zalcitabine along with reverse transcriptase inhibitors are quite effective.
- **Adverse effect**—gastrointestinal intolerance, asthenia, Headache, paresthesia, dizziness, rash, abnormal distribution of body fat, exacerbation of diabetes, crystaluria, and urinary calculi.
- **Trade name and Preparation**—Indinavir, INDIVAN, INDIVIR (400, 800 mg cap TDS), Nelfinavir—NELFIN, NEIVEX (250, 750 mg TDS), Ritonavir—RITOVIR (250, 600 mg BD)
- **Parkinsonism**—drug appears to act by promoting presynaptic synthesis and release of Dopamine in brain. It is used in milder cases, or in short course, in the dose of 100 mg bd gives good results.
- **Adverse effect**—Nausea, anorexia, insomnia, dizziness, nightmares, lack of mental concentration, hallucination, postural hypotension. Ankle edema due to local vasoconstriction.
- **Trade name**—AMANTREL, NEAMAN
- **Preparation**—Oral: 100,200 mg in elders and 5 mg/kg in children; 50 mg/5 ml syrup.
- **Precaution**—it should not be given to pregnant women and nursing mothers.

Nonselective Antiviral Agents

Interferon Alpha

They are low molecular weight glycoprotein. Interferon is produced endogenously as protective response to stimuli such as viruses, bacterial endotoxins and other intracellular microorganism. Interferon alpha and beta are produced by almost all cells against virus but interferon gamma is only produced by T-lymphocytes.

Anti-influenza Virus Agents

Amantadine

It is tricyclic amine which is unrelated to other antiviral drugs inhibit the replication of influenza A virus (A myxovirus)

- **Mechanism of action**—it has two mechanism of antiviral action. It inhibits viral replication at an early step, possibly uncoating and at late step in assembly mediated through altering hemagglutinin processing. Its target of action in influenza virus is the protein 'M₂' (integral membrane protein) acts as an ion channels. These drug interfere with the function of M₂ protein induces acid mediated dissociation of ribonucleoprotein complex in early step and potentiate acidic pH induces conformational changes in the hemagglutinin later in replication.
- **Indication:**
 - *In the prophylaxis of infection with influenza A virus*—it does not interfere with the antibody response to the influenza vaccination. So, both may be given together. If vaccine is given, amantadine can be stopped after two weeks. It is virus specific, influenza virus B is unaffected. It is given in the dose of 100 mg tablets daily for 7 days.
 - *Treatment of influenza illness*—it gives better result, if drug is given quickly just after the symptoms appear. 5 days' treatment is advised.
- **Mechanism of action**—interferon acts as immunomodulator, antiviral and antiproliferative agent.
 - Interferons are present at the site of infection before antibodies are produced, and its titre correlate with the virus act as host defense mechanism can prevent but not cures all the viral infection.
 - Interferon bind to specific cell surface receptors and affect viral replication at multiple steps like viral penetration, synthesis of viral mRNA and inhibit translation of viral protein and its release.
 - Interferon activates protein kinase enzymes and inhibits the viral protein synthesis by phosphorylation.
 - Interferon inhibits many RNA and DNA viruses, but they are host specific. Only interferon alpha has antiviral activity.
- **Indication:**
 - *Chronic hepatitis B and C*—high doses (10 MU) injected 3 cc. weekly for 6 months produce prolonged remission but relapse may occur. Addition of ribavarin to interferon alpha has the potential to further decrease the chances of relapse.
 - *AIDS related Kaposi's sarcoma*—36 million IV daily for 10-12 weeks IM or SC and maintenance dose is 36 million IV 3 times/weeks.
 - *Hairy cell leukemia*—induction dose is 3 million IV daily for 16-24 weeks SC or IM and maintenance dose is 3 million IV 3 times/weeks for 6 months.

Summary Chart						
Drugs	Trade name	Dose	Dosage form	Indications	Contraindications	Adverse effects
Idoxuridine	Ridinox	0.5 % ointment and 0.1% drops 4 hourly for 3 weeks	Ointment Drops	Keratoconjunctivitis	—	Ocular irritation , edema of lids, photophobia, bone marrow depression
Acyclovir	Zovirax Cyclovir Acivir DT	200 mg tablets for HSV infection 5 times daily for 7 days for HZV infection 800 mg 5 times daily 5% ointment-topical application 6 times daily	Tablet Injection i.v. Cream, Ointment	Genital herpes, Mucocutaneous herpes simplex, herpes zoster chickenpox, ocular keratitis	Hypersensitivity, glaucoma, Psychiatric disease Depression	Topical-stinging and burning sensation oral headache nausea malaise
Zidovudine (AZT)	Retonavir Zidovir	Adult 100, 500 mg /day in- 4 divided doses Children—180 mg/m ² 6-8 hourly. 50 mg/5 ml syrup	Capsule Tablet Syrup	AIDS	Low Hb level less than 7.5 g/deciliter, low neutrophil cell counts	Anemia, neutropenia, nausea, vomiting, abdominal pain, headache, insomnia and myalgia
Retrovir	Ritovir	250,500 mg bd	Capsule	AIDS Other AIDS related infection.	—	Rash, headache, GIT intolerance, abnormal distribution of body fat.
Amantadine	Amantrel, Neaman	Adult—100 mg bd, 200 mg OD Child— 5 mg/kg/day, 50 mg/5 ml syrup	Capsule Syrup	Prophylaxis of influenza. treatment of influenzal illness, Parkinsonism	Epilepsy, CNS disease, Pregnancy	Insomnia Dizziness Nightmares Lack of mental concentration and hallucination
Ribavirin	Ribavin Virazide	200 mg qid 50 mg/ 5ml syrup	Capsule Syrup	Influenza measles Hepatitis virus infection	—	Anemia Hemolysis, GIT symptoms
Interferon	Alferon, Realfa, viraferon	A vial (2 ml) 3,5,10 million IU/day	Injection- s.c and i.m	Chronic hepatitis. HSV, HZV, CMV, papilloma viral infection, Kaposi's sarcoma.	Severe allergies	Headache Dizziness Fever Lethargy

- *H. Simplex, H. zoster and CMV infection*—in immuno-compromised patient, it is used in the dose of 3 million IV /day or 3 times/weeks.
- Rhinoviral cold—intranasal interferon is prophylactic.
- *Condyloma acuminata caused by papillomas virus*—it is used by topical application on lesion. It can be used by an intralesional interferon injection in refractory cases.
- **Adverse effect**—
 - Flu-like symptoms: fatigue, rashes, pain, malaise, fever, visual disturbance
 - Neutropenia/ thrombocytopenia
 - Neurotoxicity: Numbness, neuropathy, tremor, sleepiness, convulsions.
 - Thyroid dysfunctions.
 - Hypotension, alopecia, liver dysfunction.
- **Trade name**—ALFERON, REALFA, VIRAFERON.
- **Preparation**—parenteral: 3,5,10 million IV vials for injections

Ribavarin

It is purine nucleoside analogue. It is active against both RNA, DNA viruses including myxoviruses, paramyxoviruses, adenoviruses, poxviruses.

- **Mechanism of action**—Ribavarin acts at multiple sites to inhibit viral replication. Ribavarin is phosphorylated intracellularly by enzyme adenosine kinase; generate its mono and triphosphate derivatives. It inhibits GTP and viral RNA synthesis.

- **Indication:**
 - *Viral bronchitis/viral pneumonia*—nebulized ribavirin is used in infants and children in the dose of 10 mg/kg/day QID. It can be administered IV and as an aerosol.
 - *Influenza*—Influenza A and B (measles in immunocompromised patient)
 - *Other disease*—it can also be given in acute hepatitis, herpes virus infection. In chronic hepatitis, it has been combined with interferon alpha.
- **Adverse effect:**
 - Anemia due to extravascular hemolysis when given orally/ intravenously.
 - Bilirubin and uric acid level are increased in plasma.
 - Long-term therapy causes CNS and GIT symptoms.
- **Trade name**—VIRAZIDE, RIBAVIN
- **Preparation**—Oral: 100, 200 mg capsules. 50 mg/5 ml syrup.
- The most commonly employed regimens are:
 - 2-NRTIs + 1-PI
 - 2-NRTIs + 1-NNRTI
 - 3-NRTIs
- Durability of these regimens depends mainly on adherence of the patient to it. Therapy should not be discontinued during an acute opportunistic infection, except in case of intolerance, interactions or toxicity.
- The 3 NRTI regimen has been less effective in patients with high (>100,000 copies of HIV-RNA/ml) initial viral loads is mostly used in less advanced cases.
- The NRTI + NNRTI PI is generally reversed for advanced disease or those with short life expectancy or repeated failures, because of risk of multiclass drug resistance and high toxicity.
- Four drug regimens have also been tested, but their superiority is not established.

Recent Advance Therapies

Highly Active Antiretroviral Therapy (HAART)

- HAART are combinations involving a protease inhibitor and other antiretroviral drugs—nucleoside reverse transcriptase inhibitors (NRTIs) and Non-nucleoside reverse transcriptase inhibitors (NNRTIs)—which have reduced the incidence of opportunistic infections.
- It has extended life substantially and decreased the infective load of HIV and other viruses.
- Orofacial disease caused by HIV infection has been significantly reduced by HAART.

Therapeutic Regimen

- Treatment should be aggressive (HAART) aiming at suppressing plasma viral load to undetectable levels (<50 copies of HIV-RNA/ml).
- Choice has to be made on the basis of efficacy, durability, drug interactions, impact on future options and cost.

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Introduction

Corticosteroids due to their wide spread of action are widely employed in medical and dental practice. The adrenal gland consists of outer cortex that secretes steroidal hormones which have glucocorticoids, mineralocorticoid and weakly androgenic activities. Glucocorticoids action is mainly for hepatic glycogen deposition, while mineralocorticoid action is for sodium, electrolyte-fluid balance.

Corticosteroid Action and its Regulation

Natural adrenocortical steroids released by adrenal cortex are controlled by pituitary release of corticotropin (ACTH). Glucocorticoids are mainly required to adapt acute stress. It takes up to 1 year following cessation of corticosteroid therapy before the Hypothalamic-Pituitary-Adrenal axis (HPA-axis) recovers fully. Its production and synthesis are tightly regulated by central nervous system.

Diurnal fluctuation in the rate of release of ACTH regulates the rate of secretion of steroids. The rate of secretion of endogenous corticosteroids is maximum in early in the morning, declines during day time and reaches a minimum at midnight as the plasma level is being highest at about 6 AM. So it is better to administer corticosteroid usually early in the morning (in between 7 to 8 PM).

Exogenous administration of corticosteroid causes suppression of normal adrenal function is the potential side effect. So, the factor that must be considered in evaluating the degree of adrenal suppression are its anti-inflammatory potency, the dosage and duration of therapy.

Dosage above 20 mg Hydrocortisone for 5 days or longer causes adrenal suppression and prevents the physiologic release of glucocorticoid. So clinician looked for 'discontinuous' steroid therapy which retains the efficacy

at lower toxicity. Small dosages of steroids at frequent interval are needed to sustain elevated plasma corticoid level, i.e. to suppress ACTH secretion.

Prolonged use of steroid preparations can result in the (HPA axis) adrenal suppression or other adverse effect. So, dentist should be concerned with the patient undergoing long term steroid therapy above the physiologic level (>20 mg/day). Short term high dose therapy, alternate day therapy and topical administration produce minimal adrenal side effect and little adrenal suppression.

In long therapy where large dosage are employed, alternate day therapy is more effective and tolerable to patient due to less side effect, as there is recovery period between each dose is equally effective. When the drug has to be used in large dosage, short acting synthetic steroids with minimal mineralocorticoid effect is to be selected.

The placenta metabolizes hydrocortisone and prednisolone to the less active cortisone and prednisone. Unlike mother the fetus cannot convert cortisone and prednisone to hydrocortisone and prednisolone. By contrast dexamethasone and betamethasone cross the placental barrier to achieve high concentrations in the fetal circulation and can suppress the fetal HPA axis. Therefore in a pregnant woman needing treatment with a glucocorticoid, prednisolone should be used in preference to dexamethasone and betamethasone.

Classification

Glucocorticoids

- Short acting [biological half life less than 12 hours]
 - Hydrocortisone (cortisol)
 - Cortisone
- Intermediate acting [biological half life in 12-36 hours]
 - Prednisolone
 - Methyl prednisolone
 - Triamcinolone

- Long acting [biological half life more than 36 hours]
 - Paramethasone
 - Dexamethasone
 - Betamethasone

Mineralocorticoids

- Deoxycorticosterone acetate (DOCA)
- Fludrocortisone
- Aldosterone

Glucocorticoids

Mechanism of Action

Cellular Action

- *Activation of receptor*—corticosteroids penetrate the cells and bind to the specific receptor protein leads to activation of receptor in the cytoplasm of cell called as 'Human-Glucocorticoid receptor' resulting from structural changes in the protein.
- *Specific protein synthesis*—this receptor moves to nucleus, interacts with specific site on chromatin and induces transcription of specific mRNA results in specific protein synthesis. This process takes at least 30-60 min. Once appropriate proteins are synthesized, effect persists for long time.

Metabolic Action

- *Carbohydrate and protein metabolism*—they prevent conversion of amino acid into proteins i.e. antianabolic effect. They promote gluconeogenesis of new glucose formation in liver. Glucocorticoids stimulate gluconeogenesis and glycogen synthesis in the fastest state. Glucocorticoids increase the serum glucose level and thus stimulate the insulin release and inhibit the uptake of glucose by muscle cells.
- *Fat metabolism*—act directly- breakdown of Triglycerides to Fatty acids (lipolysis), Indirectly-formation and storage of fats (lipogenesis). It stimulates the hormone-sensitive lipase and thus lipolysis. The increased insulin stimulate lipogenesis and inhibit lipolysis leads to net increase in fat deposition and also increases release of fatty acid and glycerol into the circulation. Prolonged administration of glucocorticoids causes a redistribution of fat in the body, with loss from extremities and a deposition in the neck (buffalo hump) and supra-clavicular area and face (moon face). Hence, the release of amino acid from muscle catabolism, supply of glucose from gluconeogenesis, inhibition of peripheral glucose uptake, stimulation of lipolysis, all are contribute to maintain adequate supply to brain and body.

- *Calcium metabolism and bone*—glucocorticoids antagonize the action of vitamin D on the gut and reduce absorption of calcium. Given in large doses for prolonged periods, they interfere with the development of cartilage and inhibit linear growth in children.
- *Electrolyte and water metabolism*—it promotes sodium retention and potassium excretion

Anti-inflammatory Action

Glucocorticoid dramatically inhibits the manifestations of inflammation viz. edema, fibrin deposition, capillary dilatation, migration of leukocytes to the inflamed region and phagocytosis.

After glucocorticoid administration, changes are maximal at 6 hours and are dissipated in 24 hours are as follows:

- *Increases in neutrophils concentration*—It is due to increased influx of neutrophils from bone marrow into the blood and decreased migration from blood vessels leading to reduction in the number of cells at the sites of inflammation.
- *Decrease in lymphocytes concentration*—T lymphocytes are decreased more as compared to the B lymphocytes. Decrease in concentration of monocytes, eosinophils, basophils due to their movement from vascular bed to lymphoid tissue.
- *Inhibition of macrophage migration factor*—it inhibits the macrophage-migration inhibition factor, tumor necrosis factor, interleukin (IL-1,IL-2,IL-3,IL-6).
- *Reduction of prostaglandin*—it influences the inflammatory response by reducing the prostaglandin, leukotriene and platelet activating factor results from activation of phospholipase and decrease expression of cyclooxygenase.
- *Vasoconstriction*—it causes vasoconstriction when applied directly to the skin by suppressing the mast cell degranulation. It also decreases the capillary permeability by reducing the amount of histamine released by basophils and mast cells.

Other Effect

- *Catabolic action*—corticoids stimulate protein and RNA synthesis. They have catabolic action on lymphoid, connective tissue, muscle fats and skin.
- *Hemopoietic action*—glucocorticoids increases the hemoglobin and red cell content of the blood, but decreases the blood lymphocytes, eosinophils, monocytes, and basophils due to their redistribution rather than destruction. It causes an increase in polymorphonuclear leukocytes and platelet count in the blood.

- *Immunosuppressive action*—glucocorticoids do not significantly decrease the concentration of antibodies in the circulation and thus the individual exhibits normal antibody-antigen response. It prevents manifestations of humoral and cellular hyperimmunity. It inhibits the development of early step of immunity by disturbing the intracellular communication among the leukocytes or function of lymphocytes. It interferes with immune mechanism by influencing the function of macrophage by inhibiting tumor necrosis factor, interleukins.
- *CNS effect*—it maintains level of sensory perception and normal level of excitability of neurons. Pharmacological doses show mild euphoria. Large dosage of glucocorticone may increase intracranial pressure.
- *Gastrointestinal effect*—it increases the basal and nocturnal gastric acid secretion. It also aggravates peptic ulcer.
- *Anti allergic*—thymus derived lymphocytes are susceptible to the actions of corticosteroids so that cell immune responses are modified following their administration. Corticosteroids, thus, in large doses can cause lysis of the T cells, prevent homograft rejection and suppress cell mediated hypersensitivity reaction.
- *Suppress release of hormone*—it suppresses the pituitary release of ACTH, GH, TSH, LH.
- *Development of fetal lung*—important effect on the development of fetal lung like production of pulmonary surface active material which is required for air-breathing is stimulated by glucocorticoids.
- *DNA synthesis*—it inhibits cell division or synthesis of DNA.
- *Topical application*—topical application is found invaluable in many dermatological, oral, ocular and external ear conditions.
- *Diagnostic test*—dexamethasone suppression of adrenal function, cortisone test in hypocalcemia and prednisolone test to distinguish intra- and extra-hepatic obstructive jaundice.
- *Dental use*—corticosteroids because of their anti-inflammatory action are used in many oral diseases. Corticosteroids are used in the management aphthous stomatitis, pemphigus, erythema multiforme, lichen planus, oral submucous fibrosis, and sinusitis.
- *Miscellaneous*—it is used in a variety of conditions like Bell's palsy, infective hepatitis, encephalitis.

Precaution

- *History*—before starting the therapy, enquire about history suggestive of peptic ulceration.
- *Glucose estimation*—examine urine periodically for sugar.
- *Weight and blood pressure record*—keep record of weight and blood pressure.
- *Do not stop abruptly*—instruct the patient not to stop the therapy abruptly.
- *Increase dose*—if the patient develops an acute infection or has to undergo therapy, increase the dose of steroids.
- *Restrict use*—glucocorticoids must not be used in the presence of infection unless the latter can be simultaneously treated with antibiotics.

Hydrocortisone (cortisol)

It is naturally occurring steroid. It acts rapidly but has short duration of action.

Uses

- *Substitution therapy*—it is used as substitution therapy in Addison's disease and hypopituitarism.
- *Intensive short term therapy*—it can save life and reduce morbidity in certain potentially lethal conditions in which inflammatory or the metabolic response of the body itself threatens life. For example, allergic emergencies such as anaphylactic shock, status asthmaticus, acute necrotizing vasculitis, etc.
- *Prolonged high dose suppressive therapy*—it is indicated in acute rheumatic fever, ulcerative colitis, subacute hepatitis, autoimmune hemolytic anemia, pemphigus, ITP, Hodgkin's disease, etc.
- *Low dose chronic palliative therapy*—use of small doses of glucocorticoids as an adjunct to some other drugs like salicylates in rheumatic arthritis. Chronic suppression of pituitary ACTH secretion is indicated in congenital, virilizing adrenocortical hyperplasia.
- *Action*—anti-allergy, anti-inflammatory.
- *Therapeutic uses*—
 - *OSMF*—hydrocortisone injected intralesionally in the areas of fibrosis in dose of 25- 50 mg/ ml fortnightly. Topically, it is applied intraorally on the oral mucosa can be combined with orabase. Hydrocortisone 25 mg tablets in dose of 100 mg/day can be given systemically helps in relieving burning sensation. Hydrocortisone 25 mg tab. Can be combined with Dexamethasone 90 mg can be given at biweekly interval. It is mainly attributed to fibrinolytic, anti-allergic, anti-inflammatory action by decreasing fibroblastic production and deposition of collagen.
 - *Aphthous ulcer*—Topical application of steroid with Orabase is effective in case of aphthous ulcer. Presence of saliva may dilute drug and effectiveness. Intralesional injections of hydrocortisone acetate (25 mg/ml) into the mucosal lesion is given.

- *Desquamative gingivitis*—topical hydrocortisone cream and ointment (0.25- 0.5%) are applied intraorally to the affected lesion gives better result.
 - *Osteoarthritis*—intraarticular injections (25 mg/ ml) are indicated in acute inflammation. Repeated injections may be associated with painless destruction of bone joint.
 - *Addison disease*—cortisol is administered 20- 30 mg / day in divided doses usually 20 mg in morning and 10 mg in afternoon as best replacement therapy.
 - *Congenital adrenal hyperplasia*—hydrocortisone is given orally as 0.6 mg / kg dose in 2-4 divided doses.
 - *Lichen planus, pemphigus, and eczematous skin disease*—for oral lesions, topical application of creams and oral suspensions, ointment 1 % applied locally twice a day. Severe episodes of pemphigus require systemic steroid administration which may be life saving drug.
 - *Ulcerative colitis, Crohn's disease*—administered orally as a lower dose on alternate day with hydrocortisone 100 mg gives better results. Care should be taken in high doses as it may result in intestinal perforation.
 - *Shock, status asthmatics*—100 mg I.V bolus + 100 mg infusion in every 8 hrs may be given.
 - *Trade name*—CORTEF, EFCORLIN, WYCORT, ORABASE-HCA.
 - *Preparation*—Oral: 5, 10, 20 mg tablets. Topical: 1% eye drop-solution, 0.025 nasal drops and 0.25-2.5% skin creams, ointment.
- Hydrocortisone acetate*—Parenteral: 25, 50 mg/ml suspension for soft tissues intralesional/intraarticular inj.
- Hydrocortisone sodium phosphate*—Parenteral: 50 mg/ml for IV, IM, SC inj.
- Hydrocortisone sodium succinate*—Parenteral: 100, 250, 500, 1000 mg / vial for IV, IM injection.

Cortisone

It is next potent drug than hydrocortisone. Nowa days it is used very occasionally.

- *Action*—anti-allergy, anti- inflammatory.
- *Therapeutic uses*—
 - *Oral submucous fibrosis*—cortisone can be given orally in dose of 25 mg tablet. Inj.25 mg/ml can be given intramuscularly, gives better results.
 - *Addison disease*—Daily dose of 25-40 mg is given as a maintenance dose.
 - *Hodgkin lymphoma*—systemic use of cortisone in lower doses can be used. Thrombocytosis /Thrombocytopenia. Drugs can be used systemically.
- *Trade name*—CORLIN, CORTONE
- *Preparation available*—oral: 5, 10, 25 mg tablets. Parenteral: 22, 25 mg/ml of solution.

Prednisolone

It is four times more potent than hydrocortisone. It is most selective glucocorticoid.

- *Action*—it is mainly anti-inflammatory but, has little sodium retentive activity. It also used as an anti-allergic and immunosuppressive.
- *Therapeutic uses*—
 - *Rheumatoid arthritis*—initial dose of prednisone 10 mg / day is given in divided doses. Initial dose should be small and increases gradually until desired degree of control is achieved.
 - *Collagen disease*—collagen diseases like Polyarteritis nodosa, Granulomatous polyarteritis. Prednisone 1 mg/kg daily is given. If favorable results are not achieved, dose is increased in 20 mg daily and then reduced gradually to 5 mg/ week.
 - *Systemic lupus erythematosus*—prednisone used intramuscularly and intravenously. For oral use, topical application of prednisone 2-3 times daily can be used.
 - *Leukemia*—mainly in acute lymphoblastic leukemia, it is used as maintenance dose. It can be used orally for at least 2-3 years.
 - *Erythema multiforme*—prednisone gives in dose of 30 mg / day for several days and terminate after symptoms subsides. It is life-saving drug in severe form of erythema multiforme.
 - *Pemphigus*—prednisone is used topically mainly for oral lesions. Intralesional injections give better results to subside oral lesions. It is used systemically to bring the disease under control. After that, the dose is reduced. Mainly prednisone with immunosuppressive drugs like cyclosporine is effective in pemphigus.
 - *Bullous pemphigoid*—systemic prednisone with immunosuppressive drugs with dapsone is given in lower doses for shorter period of time.
 - *Behçet's syndrome*—systemic prednisone with immunosuppressive drugs is used in life threatening condition. It can be used to reduce ocular, oral, genital manifestation. Intralesional drugs are administered in mucocutaneous lesions. Topical application for oral mucosal lesion which is not controlled by systemic therapy.
 - *Rheumatic arthritis*—prednisone dose of 40 mg / day in divided doses is given.
 - *Bronchial asthma*—Oral administration of prednisone is also useful. prednisone of 40-60 mg is useful to control status asthmaticus and then withdrawn gradually. Recently, inhaled corticosteroids are found for the treatment of bronchial asthma.

- *Malignancies*—prednisone is most commonly used at the dose of 30 mg/ day gives symptomatic relief in advanced malignancies.
- *Post herpetic neuralgia*—prednisone 40-60 mg given orally is used daily for 1-2 weeks. Intramuscular and intravenous injections can be given in a patient with age more than 60 years. Other diseases like amyloidosis, cyclic neutropenia, purpura, urticaria, renal diseases.
- *Trade name*—DELTA-CORTEF, PRELONE, WYSOLONE, METICORTEN.
- *Preparation available*—oral: 5, 10, 20 mg tablets. 15 mg/5 ml syrups. 5 mg/ml suspension as pediatric drops. Parenteral: Injections of 25, 50 mg/ml for soft tissue, intramuscular, intralesional, intravenously.

Methyl Prednisolone

It is slightly more potent and more selective than prednisone.

- *Action*—anti-inflammatory, antiallergic, immunosuppressive.
- *Therapeutic uses*—same as that of Prednisolone. Initial effect of methyl prednisolone is for pulse therapy. It is mainly used for rheumatoid arthritis, renal transplantation and ulcerative colitis.
- *Trade name*—MEDROL, DEPOMEDROL.
- *Preparation available*—oral: 2 mg tablets. Parenteral: 20, 40, 80 mg/ml for I.M, I.V, Intralesional, intra-articular.

Triamcinolone

It is slightly more potent than prednisolone but highly selective glucocorticoids.

- *Action*—immunosuppressive, anti-inflammatory, hemopoietic action.
- *Therapeutic uses*—
 - *Lichen planus, oral submucous fibrosis, aphthous ulcer*—injections of 0.5 to 1 ml triamcinolone acetonide (10-20 mg/ml) directly into the mucosal lesions are usually repeated at different interval. Topical application of triamcinolone 0.1% ointment is applied intraorally 2-3 times a day on mucosal lesion gives relief in several days. But presence of saliva dilutes the drug and thus reduces the effectiveness.
 - *Bronchial asthma*—triamcinolone systemic dose of 40 mg given orally. Injections of 40 mg/ml are given IM.
 - *Rheumatoid arthritis*—for relief of painful symptoms in the joint. Intraarticular injections of triamcinolone 10 to 40 mg for large joints and 2 to 6 mg/ml for small joints are administered. intralesional injections may be given for mucosal lesions.

- *Ocular disease*—ocular lesions of interstitial keratitis, spring cataract, iritis are treated by topical triamcinolone 0.1% eye drops for every 4 hours in day time.
- *Osteoarthritis*—intraarticular injections are given in the dose of 20 mg/ml.
- *Purpura*—in adults, triamcinolone 40 mg/ml is given intramuscularly until the platelet level rise to normal than tapered until the drug is withdrawn.
- *Contact cheilitis, exfoliative cheilitis, plasma cell cheilitis, granulomatous cheilitis*—triamcinolone is applied topically (0.2%) gives symptomatic relief. Repeated intralesional injections into the lips may give better result in very few weeks.
- *Benign mucous membrane pemphigoid*—topical or intralesional steroids injections are helpful.
- *Hemangioma*—intralesional and intramuscular injections of steroids are administered for better results. Injections are usually given at different intervals. It is commonly combined with immunosuppressive drugs are given in increasing frequency as it may cause hyperglycemia and osteoporosis by using single dose.
- *Trade name*—KENACORT, KENALOG, LEDERCORT, ARISTOCORT, ATOLONE
- *Preparation available*—Oral: 1, 4, 8 mg syrup. Topical: 0.1% eye drops, skin ointment. Parenteral: 3, 10, 40 mg/ml for I.M, intraarticular, intralesional injections.

Dexamethasone

It is very potent and highly selective glucocorticosteroids. It is mainly used for adrenal cortical suppression. Dexamethasone has no adverse effect on fluid retention and in hypertension.

- *Action*—immuno-inflammatory, Antiallergic
- *Therapeutic uses*—
 - *Allergic diseases, serum sickness, urticaria, hay fever, contact dermatitis, angio-neurotic edema*—dexamethasone 24 mg is administered IV. Topical application by using skin ointments on affected areas 2-4 times a day.
 - *Benign migratory glossitis*—used by topical application on affected tongue 4-5 times a day.
 - *Ocular diseases*—in case of inflammatory and allergic conditions of eyes, dexamethasone ointments, 1% eyedrops, every 4 hours in day time and at bed time.
 - *Shock, cerebral edema*—dexamethasone is mainly used in the dose of 24 mg/ml IV infusion and IM injections because this drug has fluid retention capacity. Topically can also be used.
- *Trade name*—DECADRON, DECADRON-L, WYMESONE.

- *Preparation available*—oral: 0.25, 0.5, 0.75, 1, 2, 4, 6 mg tablets. Topical: 0.1% eye drops, ear drops, skin ointments. Parenteral: 4, 8, 10, 20 mg/ml for IV, IM, intralesional and intra-articular injections. To give IV, only 24 mg/ml suspension is used. And for intraarticular injections, 16 mg/ml suspensions are used.

Betamethasone

- *Action*—this drug has similar action that of dexamethasone.
- *Therapeutic uses*—same as dexamethasone. It is mainly preferred in cerebral edema and shock.
- *Trade name*—Celestone, Betnovate, Betnesol, Betnelan.
- *Preparation available*—oral—oral drops 0.5 mg/ml, tablets 0.5-1 mg. Topical—0.1% eye drops, ointments, 0.05% nasal drops, 0.12% skin creams. Parenteral—4 mg/ml for IM, IV, intralesional and intraarticular.

Adverse Effects of Glucocorticoids

Long terms administration of glucocorticoids can cause adverse effects. These are deposition of fat in cheeks, shoulders, edema and weight gain, osteoporosis, acne, hypertension, striae/Bruises, diabetes, glycosuria, peptic ulcer, growth retardation mainly in children, delayed wound healing and adrenal suppression.

Contraindication

- *Cushing syndrome (absolute contraindication)*—most patients who are given daily doses of 100 mg of hydrocortisone of longer than 2 weeks undergo series of changes termed as cushing syndrome. It is characterized by fat deposition, rounding appearance of faces (moon facies), weight gain, muscle wasting, thinning of the skin with striae and bruising, hyperglycemia, development of osteoporosis, diabetes and impaired wound healing.
- *Diabetes mellitus*—steroid containing fluorine intensifying diabetes more than other and so it should be avoided in diabetes.
- *Hypertension*—it increases intracranial hypertension due to hypokalemia, hypochloremic alkalosis, there is increased in blood pressure.
- *Pregnancy and lactation*—it is best to avoid in 1st trimester of pregnancy due to risk of cleft palate and intrauterine growth retardation in the fetus.
- *Peptic ulcer*—risk of bleeding and silent perforation of ulcers may occur. Dyspeptic symptoms are frequent with high dose therapy.
- *Osteoporosis*—it appears to antagonize the effect of vit.D on calcium absorption and on hemopoietic system, increase in the number of platelets and red blood cells.

- *Glaucoma*—steroid increases the intraocular pressure.
- *Heart disease like CHD*—sodium retention action of corticosteroid may aggravate the CHF.
- *Herpes simplex infection*—corticosteroid are only palliative, do not remove the cause of inflammation. It favors the spread of infections as the capacity of defensive cells to kill microorganisms is impaired.

Caution

Glucocorticoids should be used cautiously in pregnancy, tuberculosis, epilepsy, in debilitating and other patients.

Newer Drugs

- *Budesonide (TN: Budecort)*—preparation available-topical 100,200,400 mcg/puff inhaler. It is used mainly in chronic bronchial asthma. Used inhalation in the dose of 200-1600 mcg/day in adults and 200-800 mcg/day in 2-4 divided doses in children.
- *Clobetasol (TN Emosone)*—preparation available-topical 0.05% ointments, lotion, gel. It is used in dermal infections by topical application on skin 2-4 times a day.
- *Fluocinolone (TN: Flucort 0.05%)*
- *Fluticasone (Flixotide 25, 50 mcg/puff inhalers)*—it is new inhaled steroid with high inflammatory activity and negligible systemic absorption. It is used as aerosol 2 times a day for prophylaxis of asthma.
- *Halocinonide (TN: Clobederm-H 0.025 to 0.1%)*
- *Mometasone (TN: ELOCON 0.1%)*—they are newer drugs used mainly for dermal infections twice a day on affected skin.

Mineralocorticoids

Important mineralocorticoids are aldosterone, fludrocortisone, desoxycortisterone.

Mechanism of Action

Mineralocorticoids have main effect on electrolyte (sodium, potassium) and fluid balance.

They have sodium retaining and potassium depleting action. Hence, it influences the salt and fluid water equilibrium in the body. Mineralocorticoids act by binding to the receptors present in the cytoplasm of principal cells forming drug-receptor complex which activate the series of events similar to described for glucocorticoids. It increases reabsorption of sodium and secretion of potassium by electrolyte and water metabolism.

Single dose of aldosterone produces sodium retention and increased urinary loss of potassium. As a result of

aldosterone deficiency the renal tubules are unable to conserve sodium.

Desoxycortisone Acetate (DOCA)

It has mainly mineralocorticoid activity. It serves as precursor of Aldosterone.

- *Action*—sodium and potassium balance retaining action.
- *Therapeutic use*—
 - *Hypoaldosteronism*—it is given systemically by intramuscular injection at weekly interval in dose of 20 mg/ml.
 - *Severe postural hypotension*—as this drug has sodium retaining action, it cures the manifestations in short period of time by using sublingual administration of DOCA in the dose of 2-5 mg/day.
 - *Addison disease*—2-5 mg sublingually, 10-20 mg intramuscularly once or twice a week is quite effective in Addison's disease. It can be given as replacement therapy.
- *Trade name*—Percorten
- *Preparation available*—Oral: 2-5 mg sublingually. Parenteral: 5, 10, 20 mg/ml IM.

Fludrocortisone

It is more potent mineralocorticoid having some glucocorticoids action.

- *Action*—sodium retention action. Anti-inflammatory action only at high doses.
- *Therapeutic uses*—
 - *Addison's disease*—0.1mg tablets are used as initial doses; gradually dose is reduced to 0.1 to 0.2 mg daily as maintenance dose. It is used to replace the Aldosterone.
 - *Autonomic neuropathy*—fludrocortisone is drug of choice for autonomic neuropathy. It is used to achieve vasoconstrictor tone. It is used in the dose of 0.5 to 1 mg/day.
 - *Hypotension*—as the drug has sodium retention capacity. It is used mainly in hypotension.
- *Congenital adrenal hypoplasia*: Tablets of 0.1 mg/day is used because in such patients, salt resting is comparatively more.
- *Trade name*—Florinef
- *Preparation available*—Oral: 0.1 mg tablets.

Adverse Effect

- Edema
- Hypokalemia
- Progressive rise in blood pressure
- Fluid and electrolyte balance disturbance

- Systemic hypotension
- Weight gain.

Contraindication

- Hypertension
- Systemic heart diseases
- Osteoporosis
- Cushing syndrome.

Dental Consideration

- *Physician consultation*—it is preferable to consult patient physician before carrying out any dental treatment in patient on steroid therapy to determine any alteration required in the dose before dental treatment.
- *Osteoporotic changes*—patient receiving long term steroid therapy results in adrenal suppression causes osteoporotic changes in jaw bone and degenerative changes in periodontium. So; dentist should look for such symptoms.
- *Stressful condition*—patient may show hypotension, vomiting, glucose intolerance, arthralgia in stressful conditions. In such conditions, majors should be taken to support blood pressure, fluid volume and to maintain normal electrolyte balance.
- *Minimum stress dental treatment*—dental treatment should be carried out with minimum stress. Minor dental procedure like minor restoration, minor biopsy, denture orthodontic adjustment, scaling and prophylaxis may not required any alteration in the dose of corticosteroids.
- *Doubling the dose*—for moderate stressful dental procedure in suppressed patients, normal dose should be doubled 8 hours before the procedure like 3rd molar impaction, vertical extraction, incision and drainage of dental infection. The dose should be tapered back to normal during post operative period after the cessation of pain, fever and other symptoms of stress.
- *Major dental surgery*—for major dental surgery under general anesthesia, due to facial trauma, severe oral infection and Orthognathic surgery, parental administration of corticoids 100 mg cortisol intramuscularly 8 hours before the procedure followed by intravenous infusion of hydrocortisone (300 mg cortisol) during procedure is given.

Suggested Reading

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Summary chart

Drugs	Trade name	Route	Dose	Indications	Contra-indications	Adverse effects
<i>Hydrocortisone</i>	Wycort (2.5%) ointment Orabase HCA (0.5%) Locoid cream (0.001%) Cortef (5 mg tablet) Aeroseb HC (0.5%) Efcorlin	Topical Topical Topical Oral Inhalation	3-4 times 3-4 times 3-4 times 100 mg 6 hourly 1-2 ml per day	Lichen planus Erythema multiforme, Pemphigus, recurrent aphthae, desquamative gingivitis, oral submucous fibrosis, Asthma, Osteoarthritis, Addison's disease	Viral infection HSV I and II Diabetes mellitus. TB, Osteoporosis, pregnancy and not to be given IV/IM	Cutaneous atrophy. Susceptibility to infection, delayed healing, glaucoma
<i>Prednisolone</i>	Predicort Prid Wysolone, Delta- cortef, meticorten, prelone	Oral	5-60 mg/day in divided doses	Lichen planus, Erythema multiforme, serum sickness, Pemphigus, thrombocytopenia, Behçet's syndrome, post herpetic neuralgia, malignancies.	Hypersensitivity, Viral infection, Diabetes mellitus, TB, peptic ulcer	Hyperglycemia, muscular weakness, psychosis, HPA axis suppression
<i>Triamcinolone</i>	Kenolog cream (0.1%) Kenolog spray (0.2%) Kenacort (1,4,8 mg) Tac-3 suspension (3 mg /ml) Aristcort suspension (25 gm/ml) Lederkort (4 mg) Lederkort ointment 0.1% Pericort-4	Topical Inhalation Oral Intralesional Intralesional	3-4 times daily 40-100 mg day 4-2 mg /day 02-03 ml/day 02-03 ml/day	Lichen planus, Erythema multiforme, recurrent aphthae, desquamative gingivitis, oral submucous fibrosis, Asthma, contact cheilitis, hemangioma.	Viral infection, TB, not to be given IV/IM	Cutaneous atrophy, Susceptibility to infection, delayed healing, Muscular weakness and psychosis
<i>Betamethasone</i>	Diprovate cream (0.05%) Valisone cream (0.1%) Betnovate cream (0.12%) Diprosone (0.1%)	Topical Topical Topical Inhalation	3-4 times daily 3-4 times daily 3-4 times daily 3-4 times daily	Lichen planus Erythema multiforme, recurrent aphthae, desquamative gingivitis, oral submucous fibrosis, Asthma, Mucous membrane pemphigoid	Viral infection Live viral immunization	Secondary infection, Delayed healing, Cutaneous atrophy
<i>Fluocinolone</i>	Flucort (0.025%) oint Cut cream (0.025%)	Topical Topical	3-4 times daily 3-4 times daily	Lichen planus, recurrent aphthae,	Viral infection	Secondary infection Delayed healing, Cutaneous atrophy

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Desensitizing Agents, Gum Paints and Mouthwashes

Dentinal Hypersensitivity

Introduction

It is certain that the teeth of man have been hurting for many thousands of years. Pain has been so closely associated with dentistry that the word “pain” and “dentistry” have almost become inseparable and one of the most important objective of dentistry has been the control and elimination of the pain.

Clinical experience suggests that dentin hypersensitivity is a relatively common cause of pain associated with the teeth. Despite this, the condition has been justifiably described as “an enigma” being frequently encountered but ill understood. Even the suitability of the term “dentin hypersensitivity” may be questioned. Such symptoms as sharp, painful, responses of short duration are associated with many conditions in which dentin is exposed including the dental caries. Furthermore, there is no evidence to indicate that “hypersensitive” dentin differs in any way from normal dentin (or that specific pulpal changes occur). The term “dentin sensitivity” may be more appropriate. The international association for the study of pain (IASP) termed hypersensitivity as “allodynia” which is pain resulting from a non-noxious stimuli to normal skin and hence, it can be appropriate to call dentinal hypersensitivity as “*allodontia*”.

Prevalence of Dentinal Hypersensitivity

- *Incidence*—the prevalence of dentinal hypersensitivity has been reported over the years in a variety of ways: Cross Sectional Surveys have shown dentinal hypersensitivity to be ranging anywhere in between 8%-57% of adult dentate population and up to 30% of adults at some time during their lifetime. *It is claimed that 14.3% of all dental patients have some degree of dentin hypersensitivity.*

- *Age group*—dentinal hypersensitivity has been shown to peak in 20 to 30 year olds and then rise again when in their 50's. The condition generally involves the facial surfaces of teeth near the cervical aspect and is very common in premolars and canines.
- *More common in periodontal patient*—72-98% of the periodontal patients suffer from dentin sensitivity due to loss of attachment and exposure of root surface. Patients undergoing periodontal treatment are particularly susceptible to this condition because of the recession following periodontal surgery or loss of cementum following non-surgical periodontal therapy.
- *More common in females*—females are more commonly affected than males as females because of esthetic reasons, females tend to brush their teeth more vigorously than males.
- *More common on facial surface*—the condition generally involves the facial surfaces of teeth near the cervical aspect and is very common in premolars, canines and second premolars.
- *More common on left side*—left side of the arch is more commonly affected than the right side of arch (Right-handed tooth brushers tend to use more force on left buccal surfaces).

Definition

Short, sharp pain arising from exposed dentin in response to thermal, evaporative, tactile, osmotic or chemical stimuli which cannot be ascribed to any other form of dental defect or pathology.

Common terminologies used for hypersensitivity are *dentin sensitivity, dentin hypersensitivity, dentinal hypersensitivity, cervical hypersensitivity/sensitivity, root hypersensitivity/ sensitivity, and cemental hypersensitivity/ sensitivity.*

Mechanism of Dentin Sensitivity

The dental pulp is richly innervated by variety of nerve fibers. (Only a few of the 1000 to 2000 nerves found in each tooth reach dentin). Pulp is richly innervated by both myelinated (A fibers -25%) and unmyelinated (C fibers -75%)—both are responsible for sensitivity of dentin. According to conduction velocity, the nerve units are classified into:

- *A fibers*—they are having conduction velocity of more than 2 m/sec. A fiber responsible for dentin hypersensitivity. They induce sharp pain in teeth.
- *C fibers*—they are having conduction velocity of less than 2 m/sec. C fibers activated only when stimulus reaches the pulp proper (intense heating). It may play a role in mediation of dull radiating pain associated with pulpal inflammation.

Causes and Clinical Features of Dentin Hypersensitivity

- *Causes*—dentin exposure due to loss of enamel from the crown of the tooth or denudation of the root surface by loss of cementum and overlying periodontal tissues. Factors responsible for it are enamel loss, occlusal wear (attrition), tooth brushing (abrasion), dietary erosion, parafunctional habit, loss of cementum, denudation of root surface (gingival recession), chronic periodontal disease, following periodontal surgery, tooth abnormally positioned in arch, root preparation, leaching margins of restorations. Improper oral hygiene technique and dental caries.
- *Symptoms* –
 - The chief symptom is a sharp, sudden pain of short duration, although some patients complain of dull, lingering sensitiveness.
 - Sensitivity to cold, but pain may also be elicited by the use of toothpick and or brushing.
 - In some cases, hot liquids and sweet or sour foods may evoke a response. Although most teeth are sensitive to more than one stimulus, not all hypersensitive teeth respond to the stimulus.

Theories of Dentinal Hypersensitivity

Transducer Theory

According to this theory the “synaptic like” relationship exists between the terminal nerve endings and the odontoblastic process.

But if true synapse were present between these two elements to facilitate the transmission of dentinal sensation then neural transmitting substance such as acetylcholine would be expected in this area of the odontoblastic process

and the predentin. There is no direct evidence for the presence of acetylcholine activity in the neural transmission in the pulp.

Modulation Theory

On any irritating stimulus to the dentin, the odontoblasts may become injured and subsequently release a variety of neurotransmitting agents as well as vasoactive and pain producing amines and proteins.

These substances may modulate associated nerve fiber action potentials by increasing neuronal clamp levels through cell membrane adenylate cyclase receptors.

Odontoblastic Transduction Theory

Odontoblasts are neural crest in origin. This theory assumed that odontoblasts extend till the periphery of the dentinal tubules and the stimuli excites the odontoblastic process.

The membrane of odontoblasts may come in close apposition with that of nerve endings in the pulp or in the dentinal tubule. These odontoblasts transmit the excitation to these nerve endings but odontoblastic process extends only part way through dentin and odontoblast membrane potential is too low to permit transduction.

Gate Control Theory and Vibration

According to this theory, the medullated nerves, i.e. A-delta fibers, can accommodate the vibrations without getting excited but unmyelinated nerve fibers, i.e. C-fibers, cannot accommodate such vibrations and get stimulated unproportionately leading to sensitivity.

Hydrodynamic Theory

It is the most widely accepted theory till date. In 1966, Branstromm provided the scientific explanation for hydrodynamic theory. Most dentinal stimuli cause pain by generating action potentials in intradental nerves through a hydrodynamic mechanism.

Application of any stimuli to dentin increases the rate of fluid flow in dentinal tubules (**Fig. 46-4**). The fluid flow in turn excites nerve terminals at the inner ends of the tubules or in the outer layer of the pulp. (Exceptions to this are electric current and intense cold, which can stimulate pulpal nerves directly).

Treatment of dentin hypersensitivity aims to interrupt this sequence of event by:

- *Creation of tubular plug*—providing a chemical ion that can react with and precipitate one of the components of the protoplasmic fluid and thus create a tubular plug.
- *Sealing of dentinal tubules*—sealing or physically occluding the outer end of dentinal tubules.

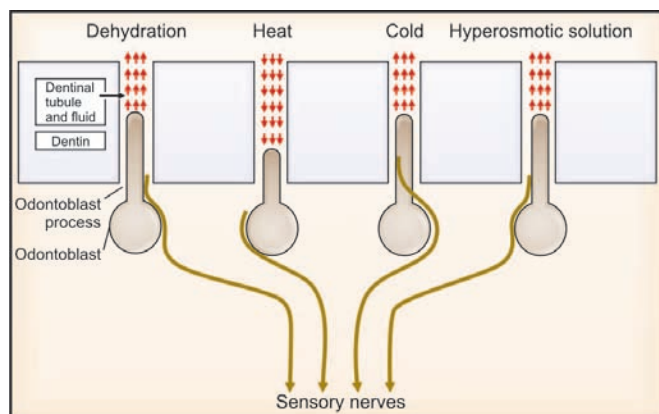


Fig. 46-1: Excitation of sensory nerve ending of dentin can cause the dentinal hypersensitivity.

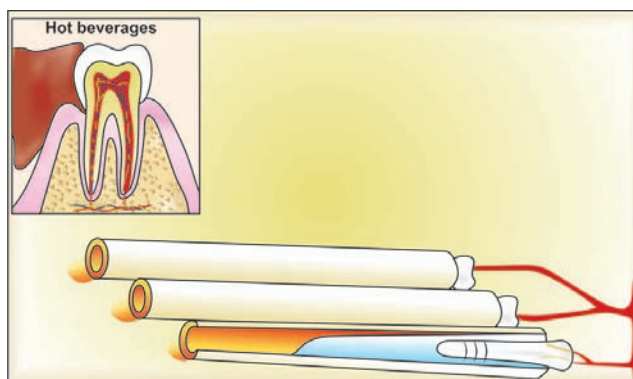


Fig. 46-2: Heat—slow inward movement of dentinal fluid.

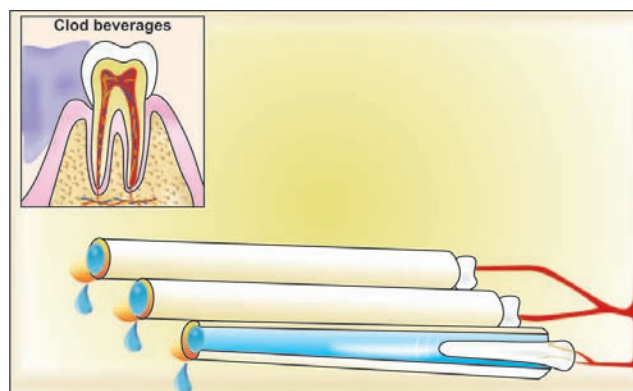


Fig. 46-3: Dehydration, cold and hypertonic solutions—outward movement of dentinal fluid.

The displacement occurs in either on outer or inward direction (**Figs 46-1 to 46-3**) and their mechanical disturbance activates the nerve endings present in the dentin or pulp.

Question to be asked for Diagnosis of Hypersensitivity

- History and nature of pain

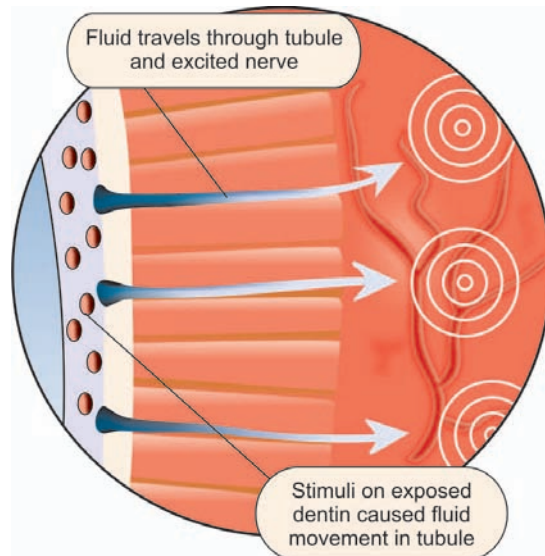


Fig. 46-4: Increase fluid flow in dentinal tubules.

- The number and location of sensitive teeth
- The area of tooth from which the sensitivity originates
- The intensity of pain
- The trigger of stimulus which initiates sensitivity
- The frequency and duration of each episode
- Recent restorative, periodontal and hygiene treatment change in diet or oral hygiene aids or home bleaching.

Differential Diagnosis of Dental Hypersensitivity

- Abscessed or non-vital tooth
- Cracked tooth syndrome
- Dental caries
- Diet sensitivity
- Genetic sensitivity
- Restorative sensitivity (Open or defective margins of restoration, fractured restorations)
- Bruxism
- Medication sensitivity
- Bleaching sensitivity
- Barodontalgia
- Gingival and periodontal pain
- Non-oral cause of dental pain like neurological causes, sinus pathology and psychogenic causes and atypical facial pain.

Clinical Examination for Diagnosis of Dental Hypersensitivity

- Tactile examination
- Air flow from air-water syringe
- Percussion
- Biting pressure

- Duration of pain after stimulus
- Radiography
- Gingival recession, loss of attachment, enamel loss
- Cracked cusps, fractured or leaking restoration, occlusal interface, hyperfunction.

Methods of Measuring Tooth Hypersensitivity

To measure the sensitivity of tooth, following methods can be used. These are mechanical, chemical, electrical, evaporative, thermal, osmotic, verbal rating scale and visual analog scale.

Mechanical (Tactile Stimuli)

- *Instrument*—it is done by explorer or probe. Constant pressure by probe (yeaple), mechanical stimulator and scaling procedure.
- *Identification of area of sensitive dentin*—all clinicians use a dental explorer to identify regions of sensitive dentin. To understand how movement of an explorer across dentin can cause a hydrodynamic stimulus, one must consider the following.
- *Applying the force*—although the use of a gentle force of 5 to 10 gm on the explorer seems as though it would be a trivial stimulus, that force is localized on the tip of explorer which is about $500 \mu\text{m}^2$. This is sufficient to overcome the elastic limit of dentin, leading not only to compression of dentin and smear layer creation under the explorer tip but also to permanent (yet microscopic) deformation of dentin (scratch development). You should lightly pass a sharp dental explorer over sensitive area of a tooth and grade the response of patient on scale
- *Results*—results of sensitivity test is as follows:
 - 0—no pain is felt
 - 1—slight pain or discomfort
 - 2—severe pain
 - 3—severe pain that lasts
- *A yeaple probe*—it is a compact handpiece that contains an explorer line in an adjustable electromagnetic field. The force should be applied to the same area at 90° to the surface in a static inwardly directed manner. The patient is asked to response whether there is pain or no pain at each test. The instrument is adjusted in 5-10 gm increments from 10-70 gm. Each increasing force compress more and more dentin. People who are more sensitive tend to react with pain at lower forces than people who are not sensitive or those who are desensitized.
- *Chemical*—it is done by hypertonic solutions like NaCl, Glucose, sucrose, CaCl_2

Electrical Stimuli

- *Instrument*—it includes electric pulp tester and dental pulp stethoscope. Pain response can be obtained from non-sensitive as well as from sensitive teeth.
- *Mechanism*—dentinal fluid movement is not necessary for transmission of electrical stimulus, rather the presence of lower resistance organic material in cementum, enamel or dentin. Sensitive teeth show lower pain thresholds than healthy teeth.
- *Technique*—electrode or probe should apply the electrical stimulus to tooth. Power source is required to vary the electrical stimulus and means of completing the electrical circuit.
- *Reference electrode*—reference electrode, a saliva ejector connecting the patient to a pulp stimulating instrument—pulp stethoscope.
- *Current applied*—electrical stimulus consisting of a direct current pulsed voltage between 0 and 150 or 0 and 300 volts.
- *Placing of probe tip*—place the probe tip on the tooth surface, depress on/off switch. Rotate the ramp adjustment dial. Intensity of electrical stimulus rises from 0 to 25 rms volts and above patient feels the pre-pain (signals the operator to remove the probe that activates the recorder).
- *Evaporative stimuli*—it includes cold air blast, air thermal system, air jet stimulator, temtronic device (microprocessor temp. Air delivery system)

Thermal Stimuli

It is electronic threshold measurement device, cold water testing and heat.

- *Why cold stimuli is used*—because patient are generally more sensitive to cold than to hot stimuli, the use of cold water (10, 15, 20, 25, 30°C) as a simple, quantitative stimulus is gaining in popularity.
- *Mechanism*—thermal stimuli are effective of differences in thermal conductivity and coefficient of expansion or contraction of fluids and their container, enamel and dentin. Thus application of cold causes more rapid volumetric contraction of dentinal fluid that occurs in the dentin. This mismatch of volumetric changes produces negative intrapulpal and intradental pressures that displace mechanoreceptors and cause pain. Heating has the opposite effect but the same result, pain clinically, cold stimuli are more useful than hot stimuli for testing dentinal sensitivity. Patients tolerate cold stimuli better than hot stimuli, and there is danger of causing pulpal damage.
- *Isolation of tooth*—in using cold water, each tooth is isolated with a rubber dam and water at a known temperature.

- *Flowing of water*—water is slowly flowed on the exposed dentin surface for a maximum for 3 seconds from a disposable plastic syringe.
- *Reaction of patient*—the patient is forced to decide if that temperature causes pain or not and then the next lower temperature. It is tried until the patient responds unequivocally.
- *Air burst method*—another method which can be used is directing a burst of air (room temperature) from a dental syringe onto the tooth to be tested. Room temperature is cooler than the tooth and cooling by this method can be easily detected as pain if the teeth are sensitive or parallel. 1 second blast from the air syringe, temperature between 65 and 70°C and pressure of 60 psi at right angle to the tooth near CEJ or exposed root surface is used.

Osmotic

It is subjective pain to a sweet stimulus.

- *Solution used*—fresh saturated solution of sucrose; allowing it to reach room temperature.
- *Application*—a cotton applicator saturated with the solution applied to the root surface, allowed to remain for 10 seconds.
- *Results*— no pain—0
and pain –1.

Verbal Rating Scale (VRS)

It presents a restrictive choice of words that may not represent the pain experience with significant precision for all patients (Huskinson 1974). Keel [1948] described a four point scale grading pain as slight, moderate, severe and agonizing. More recently, modification of this type of pain scale has been reported and are as follows:

- *Hansen 1992*—simple binary pain scale- pain before the treatment/no pain after the treatment.
- *Gilliam and Newman 1993*—it is as follows
 - 0 = no discomfort
 - 1 = mild discomfort
 - 2 = marked discomfort
 - 3 = marked discomfort that lasted 10 sec.
- *Gedalia et al 1987*
 - 1 = no pain
 - 2 = discomfort only
 - 3 = pain
 - 4 = severe pain
 - 5 = unbearable pain
- *Thrash et al 1992*—
 - 0 = no significant discomfort
 - 1 = discomfort, but no severe pain
 - 2 = severe pain during application of stimulus
 - 3 = severe pain during application and continuing after application of stimulus.

- *Schiff et al 1998* (Schiff's cold air score)
 - 0 = tooth/ subject does not responds to air stimulus
 - 1 = tooth/ subject responds to air stimulus but does not request discontinuation of air
 - 2 = tooth/ subject responds to air stimulus and requests for discontinuation of air stimulus or moves away from stimulus.
 - 3 = tooth/ subject responds to air stimulus considers stimulus to be painful and requests for discontinuation of stimulus.

Visual Analog Scale (VAS)

A Visual Analogue Scale (VAS) is a measurement instrument that tries to measure a characteristic or attitude that is believed to range across a continuum of values from none to an extreme amount of pain.

Method

Ask the patient to indicate on the line where the pain is in relation to the two extremes. Ask the patient to mark on the line the point that they feel represents their perception of their current state (**Fig. 46-5**). The VAS score is determined by measuring in millimeters from the left hand end of the line to the point that the patient marks. When the VAS is properly explained to subjects, they can easily understand its use and successfully use it to indicate their level of pain response to hypersensitive stimuli.

- *Comparison between VAS and VRS*—the VAS should be the more appropriate device than the VRS for measuring levels of sensitivity, pain during subject assessment and for measuring tactile and thermal stimuli of hypersensitivity. For those unable to read or to understand the linear visual analogue scale, picture of faces showing increasing distress can be used (**Fig. 46-5**).
- *Advantage of VAS*—VAS is found to be reproducible therefore a very high correlation between successive measurements of pain severity has been noted.

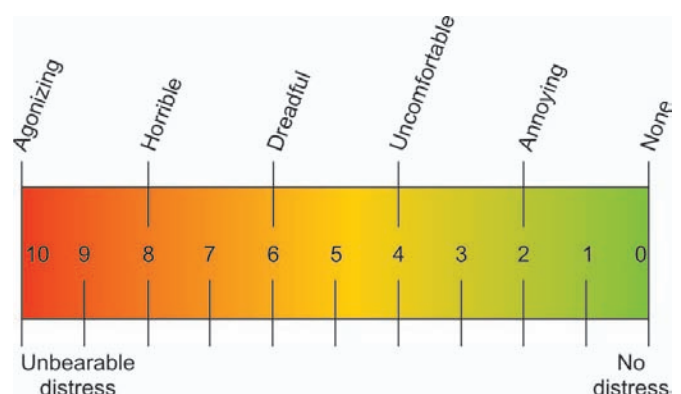


Fig. 46-5: Visual rating scale is used to determine current state.

- *Disadvantage*—disadvantage of visual analog scale is that patient tends to spread their responses over the entire scale regardless of the magnitude of the actual sensation.

Treatment of Hypersensitive Dentin

Treating dentinal hypersensitivity can be challenging for the dental professional because of the difficulty related to measuring the pain response since the response varies from patient to patient. In addition if the dentin exposure is due to personal habits, it may be difficult for patients to change their behavior(s). If the diagnosis confirms dentinal hypersensitivity in the absence of underlying diseases or structural problems, then the following steps can be initiated:

- Remove the risk factors by educating the patient about dietary acids and other oral care habits;
- Recommend different toothbrushing methods, if appropriate;
- Initiate treatment by recommending a desensitizing agent for home use; or
- Applying topical desensitizing agents professionally. Grossmann's ideal requisites for a desensitizing agent.
 - *Non irritant*—it should be non-irritant to the pulp.
 - *Painless*—it should be relatively painless on applications
 - *Easy application*—application of desensitizing agent should be easy.
 - *Action*—action should be rapid.
 - *Long term effect*—effect of desensitizing agent should be long term.
 - *No staining*—no staining of tooth should not occur.
 - *Consistency*—desensitizing agent should be consistent in effectiveness.

Traditional Treatment of Dentinal Hypersensitivity

Home care with dentifrices

- *Potassium nitrate dentifrices*—Thermoseal, Thermoseal RA, Thermokind-f, Sensicure k, Sensodent-k, Sensodent-kf, Promise, S
- *Senquel-f, Nitra, Denquel, Sensodyne.*
- *Potassium oxalate dentifrice*—Protect.
- *Sodium fluoride dentifrices*—Thermodont, Thermoseal, Colgate, Thermodont, Proevident.
- *Sodium citrate dentifrices*—Protect
- *Sodium monofluorophosphate dentifrices*—Colgate, Thermokind-f, Senquel-f, Sensicure, Nitra, Sensicure k, Aquafresh, Pepsodent, Sensodent-k, Sensodent-r.
- *Stannous fluoride dentifrice*—Colgate gel, Colgate flourigard, Cibacca fluoride
- *Calcium chloride*—Forhan's
- *Calcium carbonate*—Colgate, Meswak, Neem active tooth paste

- *Strontium chloride*—Thermoseal, Sensodyne, Thermodont, Senolin, Stolin, Sensicure
- *Triclosan*—Thermokind-f, Sensicure, Pepsodent, Colgate, Senquel-f
- *Zinc sulfate*—Sensicure, Sensicure k, Sensodent-r

In-Office treatment (professional application in dental office)

- *Physical agents*—composites, resins, varnishes, sealants, soft tissue grafts, GIC (glass ionomer cement), and laser sealing off tubules.
- *Chemical agents*—corticosteroids, silver nitrate, zinc chloride, strontium chloride, formaldehyde, calcium hydroxide, potassium nitrate, potassium oxalate, fluorides, sodium citrate, sodium monofluorophosphate.

Mechanism of Action of Desensitizing Agents

Most of the therapies proposed till date for treatment rely on one of the two major suppressive mechanisms, i.e., sealing off the dentinal tubules or dampening neural impulses (potassium nitrate).

Sealing off or the reduction in the diameter of the tubule so as to limit the displacement/ flow of fluid in the dentinal tubules can be achieved by

- *Formation of smear layer*—it is produced by burnishing the exposed root surface. (orange woodstick or toothpick—partially occlude to dentinal tubule)
- *Formation of insoluble precipitates*—topical application will form insoluble precipitates within tubules. for example-
 - *Oxalate*—oxalate from potassium oxalate reacts with the ionized calcium in the dentinal tubule and forms insoluble calcium oxalate crystals. The crystals block the dentinal tubules and prevent fluid flow through the tubule.
 - *Silver nitrate*—silver nitrate precipitates protein constituents of odontoblastic processes, thereby partially blocking the dentinal tubules.
 - *Sodium fluoride*—sodium fluoride reacts with calcium and phosphate ions and forms calcium fluoride crystals thus blocking the dentinal tubule
 - *Stannous fluoride*—stannous fluoride forms dense layer of tin and fluoride containing tubular particles thus effectively blocking the dentinal tubules.
 - *Fluoride iontophoresis*—iontophoresis act by influencing ionic motion by electric currents which may enhance ion uptake by the dentinal tubules and aid in achieving desensitization. The process of influencing ionic motion by electric currents is known as electrophoresis, cataphoresis, or iontophoresis. The objective of fluoride iontophoresis is to drive fluoride ions more deeply into the dentinal tubule that cannot be achieved with topical application of fluoride alone.

- *Impregnation of tubules with resins*—sealing off the tubules with plastic resins: glass ionomer cement.
- *Sealing by dentin bonding agents*—sealing off the dentinal tubule by dentin bonding agents. They seal the dentinal tubules and thus prevent pain producing stimuli from reaching the pulp. Procedure is as follows:
 - Clean the sensitive dentin—each for 5 sec—wash and dry for 15-20 sec
 - A drop of enamel bond applied to dentin
 - Gluma—dentin bonding agent—5% glutaraldehyde primer and 35% hema (hydroxyl ethyl methacrylate).
 - Immediate and strong attachment to dentin
 - Prevent bacterial overgrowth in tooth/restoration interface—beneficial effect in inhibiting plaque accumulation on sensitive root surface
- *Surgical root coverage procedures*—various procedures are used to cover the denuded areas of teeth such as free gingival graft, connective tissue graft, laterally positioned flap, coronally positioned flap, subepithelial connective tissue graft and guided tissue regeneration.
- *Laser sealing off tubules*—helium-neon (HeNe) and Nd:YAG lasers are found to be effective in the treatment of dentinal hypersensitivity without detrimental pulpal effects. The lasers used for the treatment of dentin hypersensitivity can be divided in two groups.
 - *Low output power lasers*: helium-neon (HeNe) and gallium/aluminium/arsenide (Ga-Al-As) lasers.
 - *Middle output power lasers*- Nd: YAG (1064 nm) and CO₂ lasers (10,600 nm) at different wavelengths and exposure time are used to block the dentinal tubules effectively.
- Avoid using large amounts of dentifrice or reapplying additional dentifrice during brushing.
- Avoid hard bristled tooth brushes.
- Avoid over brushing with excessive pressure for prolonged periods of time.
- Avoid excessive flossing or incorrect use of other interproximal cleaning devices.
- Avoid picking at the gum or using tooth picks inappropriately.

Suggestions for Professionals

- Avoid over instrumentation of the root surfaces during calculus removal and scaling and root planing as it may cause
- Avoid over polishing the exposed roots during stain removal.
- Avoid violating the biologic width when placing crown margins causing subsequent recession.
- Avoid burning the gingival tissue during in-office tooth whitening or bleaching procedures.

Summary and Conclusion (Table 46-1)

Dentinal hypersensitivity is one of the most painful and least predictably treated chronic conditions in dentistry. In clinical practice, the professional approach to dentin hypersensitivity has been heavily treatment-based with little regard for control of the etiological agent and predisposing factors, which creates the problem. This is not surprising since the dentist and the sufferer are virtually bombarded with a vast array of products formulated to treat dentin hypersensitivity.

Patients treated for dentin hypersensitivity should be counseled about dietary acids and the importance of proper effective oral hygiene. Over-the-counter dentifrices will continue to have an important role in treating dentin hypersensitivity. Potassium nitrate, potassium oxalate, iontophoresis with 2% sodium fluoride is proven to be effective with varying degree of success in reducing the dentin hypersensitivity. In the future, dental lasers are expected to have an important role in treating hypersensitivity.

Instructions to be given to Patients while Prescribing Desensitizing Agent

- Hypersensitivity appears as a result of exposure of dentin, which is inevitable if calculus and plaque and their products are to be removed.
- Hypersensitivity slowly disappears over a few weeks.
- Plaque control is important for the reduction of hypersensitivity. Patient is instructed about proper tooth brushing technique as improper tooth brushing is one of the etiologic factors in dentin hypersensitivity.
- Desensitizing agents do not produce immediate relief. They must be used for several days to weeks (2-6 week) to produce results.

Prevention of Dentin Hypersensitivity

Suggestions for Patients

- Avoid gingival recession due to poor plaque removal by practicing good oral hygiene technique.

Mouthwashes (Table 46-2)

Mouthwashes are usually aqueous solutions in concentrated form of a substance with deodorant, antiseptic, local analgesic and astringent properties.

Mouthwashes comprise a large group of liquid compound used as oral rinse for cosmetic or therapeutic purpose.

Table 46-1: Desensitizing agents

Desensitizing agents	Trade name of toothpaste	Mechanism of action
Strontium chloride	Thermoseal Eanisam Desent Senolin Toss	Bicollodial binding and blocking of micro-tubular fibrillae on the exposed dentin. Strontium ions stimulate secondary dentin formation Modification of transmission of impulses and stimulation for re-calcification
Potassium nitrate	Senquel Sensodent K	Oxidizing effect or blocking of dentinal tubules
Formalin	Sensoform Dentoform	Protein precipitation
Strontium chloride and formaldehyde	Stolin	Same as strontium chloride and formalin
Potassium nitrate 5% sodium mono-fluorophosphates 0.7% available fluoride	Senquel-F	

Table 46-2: Mouthwashes

Drug	Trade name
0.2 % chlorhexidine gluconate	Clohex, hexidine, rexidin
0.12% chlorhexidine and 2% sodium benzoate	A.M-P.M.
1% Povidone iodine, essential oils	Betadine, alphasine, povidax, povidone
0.15% benzydamine hydrochloride	Tantum oral rinse
1.02% chloroxylenol and 0.12% menthol	Dettolin
Chamomilla extract in alcohol (42.8%), mentha oil (18.5 mg) anise oil (7 mg) and alcohol (0.1ml)	Kamillosan-N
Thymol (0.06%), eucalyptol (0.9%), benzoic acid (0.15%), menthol (0.04%)	Listerine

Indications for Antiplaque Mouthwash

- *Replacement for mechanical tooth brushing*—antiplaque mouthwash is used to replace mechanical tooth brushing when this is not possible acute oral mucosal and gingival infections, after periodontal or oral surgery, during the healing period, after cosmetic jaw surgery, in case of patient using intermaxillary fixation, and for mentally and physically handicapped patients.
- *Adjunct use*—as an adjunct to normal mechanical brushing in situations where this may be compromised by discomfort or inadequacies.
- After scaling when there is cervical hypersensitivity due to exposed root surfaces, prescribe mouthwashes for about 4 weeks. Measures to treat hypersensitivity should also be instituted simultaneously.
- Following subgingival scaling and root planning when the gingiva may be sore for a few days, use of a mouthwash is recommended for about 3 days.
- *Cleaning of mouth*—vigorous mouthwashing does help in keeping the mouth clean by moving the food debris, mucus and even bacteria.
- *Decrease bacterial population*—it is also used to decrease bacterial population, to provide topical anesthesia.
- *Reduction of oral acidity*—it also reduces oral acidity and inactivates odor producing salivary or bacterial enzymes.
- *Refresh the mouth*—the patient may use mouthwashes to debride the oral cavity after tooth brushing and to refresh the mouth or as an attempt to correct halitosis. From simple breath fresheners to products that can really influence oral health, a variety of mouthwashes are available in the market.
- *Resolution of chronic gingivitis*—Antiplaque or antimicrobial mouthwash is used to inhibit bacterial plaque formation and prevent or resolve chronic gingivitis. They can affect only supragingival plaque. So they have no role in the treatment of existing periodontal disease, since

they cannot either reach the subgingival environment or penetrate thick layers of established plaque. In these situations, they are used after supra- and subgingival scaling has been done, rendering the tooth surfaces clean, in order to maintain this situation for a short period when the soreness of the gingiva may prevent effective mechanical plaque control.

- *Indication for fluoride containing mouth rinse*—it is used to prevent dental decay. They may be recommended for children having orthodontic treatment, children with high caries risk, dry mouth and after radiation therapy.

Types of Antiplaque Mouthwashes

- *Mouthwashes containing essential oils*—essential oils derived by distillation from plants, have a characteristic odor and taste and should not be used in a concentration greater than 0.2%. The most commonly used essential oils are eucalyptus oil, methylsalicylates, orange oil, peppermint oil and spearmint oil. Listerine, one of the oldest mouthwashes available, is an essential oil/phenolic mouthwash. It has been shown to have moderate plaque inhibitory effect and some anti-gingivitis effect. Its lack of profound plaque inhibitory effect is because it has poor oral retention. Ethyl alcohol is used to enhance the solubility of organic compounds in water and should not exceed a concentration of 10% because of possible local irritation.
- *Phenol and its derivative*—they are also used for their antibacterial and anodyne effects in mouthwash preparations. Their value is limited by their toxicity, objectionable taste, sensitization properties, high cost and decreased activity in the presence of organic matter. Commonly used phenol derivatives are hexylresorcinol and thymol.
- *Mouthwash containing oxygenating agents*—oxygenating agents like Hydrogen peroxide, buffered Sodium peroxyborate and peroxy carbonate in mouthwashes have a beneficial effect on acute ulcerative gingivitis, probably by inhibiting anaerobic bacteria. Mechanism by which they inhibit anaerobic bacteria is that they release molecular oxygen which can destroy anaerobic bacteria.
- *Mouthwash containing bisguanide antiseptic*—bisguanide antiseptics, like Chlorhexidine, Alexidine and octenidine possess antiplaque activity. These are cationic agents with fungicidal activity and bactericidal action against gram positive and gram negative organisms. Bisguanide antiseptics are able to kill a wide range of micro-organisms by damaging the cell wall.
- *Mouthwash containing triclosan*—triclosan, a trichloro-2'-hydroxy diphenyl ether, is a non-ionic antiseptic. It has a moderate antiseptic effect when used as a mouthwash

in combination with zinc. It has been shown to reduce histamine induced dermal inflammation and reduces the severity and healing period of aphthous ulcers.

- *Colgate total plax mouthwash*—colgate Total Plax mouthwash has Triclosan and Sodium fluoride as its components. Triclosan has little or no substantivity, but its oral retention can be increased by its combination with copolymers of methoxy ethylene and maleic acid.
- *Povidone iodine*—povidone iodine appears to have no significant plaque inhibitory activity when used as 1% mouthwash and the absorption of significant levels of iodine through the oral mucosal may make this compound for prolonged use in the oral cavity. It could cause problem of iodine sensitivity in sensitized individuals. Piodin (Glaxo Wellcome), povidine Gargle (Stadmed) are povidone iodine mouthwashes available in the market.
- *Mouthwash containing sanguinarine*—it is a component of an alkaloid extract obtained from the dried rhizome of the bloodroot plant, sanguinaria Canadensis. Recently, it is incorporated with zinc chloride in dentifrices and in mouthwashes as an antiplaque and anti-gingivitis agent. The antibacterial activity is due to its ability to inhibit sulfhydryl dependent enzymes.

Alcohol Content of Mouthwashes

Most mouthwashes contain pharmaceutical grade alcohol, as a preservative and as a semi-active ingredient. Significant amounts of alcohol contained in many mouthwashes can lead to certain disadvantages. Care should be taken that they are not accidentally swallowed, especially by children, to avoid toxicity. Small children should not be advised mouthwashes, because they are not able to spit out properly. Moreover, most children have good gingival health.

Because of known links between alcohol consumption plus tobacco smoking and oral and pharyngeal cancer, it has been suggested that the frequent use of alcohol containing mouthwashes might increase the incidence of this form of cancer.

Lastly, alcohol containing mouthwashes have been shown to reduce the hardness of composite and hybrid resin restorations

Chlorhexidine Mouthwash

Chlorhexidine molecule gets adsorbed onto the oral surfaces and gets released at bactericidal level over prolonged periods. Due to this process, Chlorhexidine has antiplaque properties unsurpassed by other agents. The antibacterial action of Chlorhexidine is due to an increase in cellular membrane permeability followed by coagulation of the cytoplasmic macromolecules. It is effective *in vitro*

against Gram +ve and Gram -ve bacteria including aerobes and anaerobes and yeast and fungi.

The substantivity (the ability of drugs to adsorb onto and bind to soft and hard tissues) of Chlorhexidine was first described in the 1970s. Due to this property, Chlorhexidine can maintain effective concentration for prolonged periods of time. Different brands of Chlorhexidine are available in the market, e.g., Rexidin (Warren), Clohex (Group) and A.M.-P.M (Elder).

Structure

Chlorhexidine is a chlorophenyl bisguanide that has been used as an acetate and more commonly as a gluconate salt. It is N, N-bis (4-chlorophenyl)-3, 12-dimino-2, 4, 11, 13-tetra-azotetradecanedimidamide di-D-gluconate.

Mechanisms of Action

- *Absorption onto oral surface*—chlorhexidine molecule gets adsorbed onto the oral surfaces and is released at bactericidal levels over prolonged periods. Due to this process, chlorhexidine has antiplaque properties unsurpassed by other agents.
- *Antibacterial action*—this action of chlorhexidine is due to increase in cellular membrane permeability followed by coagulation of the cytoplasmic macromolecules. It is effective *in vitro* against gram +ve and gram -ve bacteria including aerobes and anaerobes and yeast and fungi. The positively charged chlorhexidine binds to negatively charged microbial cell surface. It is followed by disorganization of cytoplasmic membrane. Low concentration allows cytoplasmic constituent to leak out and high concentration coagulates them.
- *Blocking of acidic groups*—these groups on the salivary glycoprotein reduce the protein absorption to tooth surfaces.

- *Cell wall penetration*—in high concentration, chlorhexidine penetrates cell wall and causes precipitation of the cytoplasm.

Indications

- *Periodontal disease*—it is used in all forms of periodontal diseases.
- *Subgingival irrigation*—it is also used for sub-gingival irrigation of periodontal pockets.
- *Aphthous ulcer*—it also reduces the severity and duration of aphthous ulceration.
- *Substitution of mechanical plaque control*—where mechanical plaque control is not useful.
- *Denture stomatitis*—denture stomatitis has been treated by soaking dentures in 0.2% chlorhexidine overnight for 5 months combined with daily use of amphotericin B lozenges for 14 days.

Side Effects of Chlorhexidine

- *Unpleasant taste*—it has an unpleasant taste and it alters taste sensation. Interference with taste sensation is caused by denaturation of surface proteins on the taste buds.
- *Stains*—it produces brown stains on teeth, which is very difficult to remove. This can also affect the mucous membranes and tongue and may be related to the precipitation of chromogenic dietary factors onto the teeth and mucous membranes. Due to this reason, it is important to advise patients using chlorhexidine mouthwash to avoid the intake of tea, coffee and red wine during the duration of its use. Remember to severely restrict its use in patients with visible anterior composite and glass ionomer restorations since they also get stained.
- *Calculus formation*—chlorhexidine encourages supra-gingival calculus formation. It may be due to rise in pH

Table 46-3: Gum paints

Drugs	Trade name
2% zinc sulfate	Gingisol
Tannic acid (66.27%), glycerin, thymol (0.033%), potassium iodide (0.05%), menthol (0.05%) and iodine (0.03%)	Sensoform
Choline salicylate (8.7%), cetalkonium chloride (0.01%) and ethyl alcohol (39%)	Gelora
Choline salicylate (9%) and benzalkonium chloride (0.02%)	Zytee
Iodine (1gm), potassium iodide (2gm), alum (1.5gm), tannic acid (0.5gm), thymol (0.25mg), camphor (0.2mg) and menthol	Toss gum paint
Choline salicylate (8.7%), lignocaine hydrochloride (2%) and benzalkonium chloride solution (0.01%)	Dentogel Dologel
Tannic acid (2%), zinc chloride (1%) and cetrimide (0.1%)	Stolin

as a result of absorption of cationic agents and marked impact on bacterial integrity.

- *Soreness of oral mucosa*—desquamation and soreness of the oral mucosa can occur. It is due to precipitation of mucin layer, by reducing its lubricating effect.
- *Transient parotitis*—transient parotitis has also reported.
- *Altered taste sensation*—chlorhexidine mouthwash may alters taste sensation.

Gum Paints (Table 46-3)

Gum paint is just a stronger solution of a drug, which is used as mouthwash. They are used for topical application. They develop a strong concentration of the drug against inflamed tissue. They are combination of antiseptics and tanning agents. They are used for local application to the gums during severe bacterial infection and their inflammatory conditions. To apply it you have to dry the area and apply using cotton or index finger and it is not to be swallowed. When applied they provide a cooling, soothing, astringent effect and they have germicidal, fungicidal, anesthetics and healing properties. It is used in the treatment of stomatitis, inflamed/bleeding/spongy and painful gum conditions. It reduces sensitivity and increase gingival resistance against infection.

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Introduction

Administration of drugs to pregnant patient is of significant concern. Two main concerns must be addressed when considering whether to give a drug to pregnant women. The first is that the drug may be teratogenic and the second is that drug can affect near term fetus.

One must always be aware of the teratogenic, toxic or otherwise harmful effects of the drug on the developing fetus. The physiologic changes during pregnancy and the consequent alteration in the pharmacokinetics lead to changes in drug absorption, distribution, metabolism and excretion.

As a general rule, it is best that no drug should be given during pregnancy, especially during the first trimester as it is period of organogenesis. Fortunately, most of drugs commonly used in dentistry are not contraindicated during pregnancy. Tetracycline and streptomycin are notably exceptions.

The fetus may metabolize a particular drug via a different pathway from that of mother or its metabolic product may bound more avidly in fetal thus leading to accumulation of drug or its metabolites in fetus. Also since body systems of fetus are not fully developed, the fetus cannot process medicine like mother's system so same drug can cause harm to fetus.

Pregnancy Trimesters

Pregnancy involves three trimesters each 3 months long.

- *First trimester*—in this trimester, different body organs in the fetus are forming. It is most critical time for teratogenicity. Dental prophylaxis with detailed instructions and a visual examination of the oral cavity without X-rays should be performed if the patient is pregnant. Elective dental treatment should be avoided

in the morning as women may feel nauseated in the morning.

- *Second trimester*—it is an excellent time for the patient to undergo dental prophylaxis if needed. The patient's periodontal status should be carefully evaluated during this period.
- *Third trimester*—the women begin to feel uncomfortable and it is difficult for her to lie in prone position for long period time. Drugs that may affect the newborn should not be given during this trimester. Positioning of patient on dental chair can cause hypotension due to compression of gravid uterus on the inferior vena cava, resulting in syncope. Stress can precipitate premature labor. Due to hormonal changes, gingival tissue shows exaggerated response to local irritants.

Pharmacokinetics in Pregnancy

- *Drug absorption*—high circulating levels of progesterone slow the gastric emptying as well as gut motility resulting in slower drug absorption. Parenteral drug administration is preferred in order to obtain a quick response. Drug compliance may be poor because of nausea and fear of adverse effect.
- *Drug metabolism*—hepatic drug metabolizing enzymes are induced during pregnancy probably by high concentration of circulating progesterone. This can lead to more rapid metabolic degradation especially of highly lipid soluble drugs. However, this is of little clinical consequence.
- *Drug excretion*—during pregnancy the renal plasma flow increases by 100% and glomerular filtration rate by 70%. Hence, drugs which depend for their elimination mainly on kidney are eliminated more rapidly than in non-pregnant stage, e.g. ampicillin, gentamicin and cephalosporin.

Recommended Drugs to be used in Pregnancy				
Type	Drugs	1st Trimester	2nd and 3rd Trimester	Comment
Local anesthetics	Lidocaine	Yes	Yes	First choice anesthetics, fetal bradycardia near term
	Mepivacaine	Yes	Yes	Fetal bradycardia near term
	Bupivacaine	No	No	Embryocidal in rabbits and high lipid solubility
	Benzocaine	Yes	Yes	Fetal bradycardia near term
Vasoconstrictors	Epinephrine	Yes	Yes	It can produce hypoxia (use cardiac dose)
Analgesics	Paracetamol	Yes	Yes	Teratogenic at over dose level
	Codeine	Limited dose	Limited dose	Respiratory distress near term high dose contraindicated
	Aspirin	No	No	Bleeding, prolonged parturition, premature closure of patent ductus arteriosus
	Ibuprofen	Yes (cautiously)	No	Same as aspirin
Antibiotics	Penicillin	Yes	Yes	Safe
	Erythromycin	Yes	Yes	Safe except estolate form
	Tetracycline	No	No	Stains teeth, affects bone
	Clindamycin	No	No	Only if alternative dose not exist
	Cephalosporin	Yes	Yes	Safe only if use as indicated
	Metronidazole	No	No	Carcinogenic and mutagenic in animals
Antifungal	Clotrimazole	No	Yes with caution	Poorly absorbed following topical or intravaginal application, abnormal liver function test in adults can occur.
	Ketoconazole	No	No	Embryotoxic in rat
	Nystatin	Yes	Yes	Safe
Sedative	Benzodiazepines	No	No	Cleft lip, neural tube defect
	N ₂ O with 50% O ₂	No	Possible with adequate O ₂	Ensure adequate oxygen intake, female operators avoid chronic exposure

Use of dental drugs by nursing mother			
Types	Drugs	Acceptable	Watch infant for symptoms of
Local anesthetics	Amides	Yes	Central nervous system changes
Vasoconstrictors	Epinephrine	Yes	Hyperactivity or irritability
Analgesics	Aspirin	Yes with caution	Avoid feeding for 1 hour after dose, occasional low dose pose no hazard, chronic high dose may pose problems
	NSAID	Yes with caution	Use ibuprofen (concentration in milk is low) avoid long acting NSAIDs
	Acetaminophen	Yes	Present in milk in small amount (peak 1-2 hours)
	Opioids	Yes	Small doses no problems and in larger doses sedation, poor feeding and constipation
Antibiotics	Penicillin	Yes	Allergic symptoms, diarrhea
	Erythromycin	Yes	Present in milk, diarrhea
	Cephalosporin	Yes	Allergic symptoms, diarrhea
	Tetracycline	No	Tooth staining
	Clindamycin	Yes/no	Diarrhea, pseudomembranous colitis
	Metronidazole	No	Carcinogenic in animal
Antifungal	Nystatin	Yes	Not absorbed into systemic circulation from mouth or gastrointestinal tract
	Clotrimazole	Yes	Excreted in milk, use nystatin first
	Ketoconazole	No	Express and discard milk
Antiviral	Acyclovir	Yes	Concentrated in milk
Antianxiety	Nitrous oxide	Yes	Excreted through mother's lung
	Benzodiazepines	No	Sedation, infants metabolize oxidized agent more slowly

- *Increase total blood volume*—there is increased total blood volume, because of increased fluid retention. This leads to change in cardiac output, blood pressure and glomerular filtration rate. This results in change in volume of distribution of drug, change in metabolism, change in absorption, change in excretion of drug, change in protein binding of drugs and passage of drug through placenta.
- *Teratogenicity*—it refers to capacity of a drug to cause fetal abnormalities when administered to pregnant mother. Drug can affect fetus at three stages, i.e. stage of fertilization and implantation, stage of organogenesis and stage of growth and development

Drugs	Abnormalities
Thalidomide	Phocomelia
Anticancer drug	Multiple defect, Fetal death
Tetracycline	Discolored and deformed tooth, Retarded bone growth.
Phenytoin	Craniofacial and limb defect, cleft lip, cleft palate
Phenobarbitone	Various malformations
Carbamazepine	CNS defect
Retinoids	Various abnormalities
Alcohol	Fetal alcohol embryopathy

Dental Management of Pregnant Patient

- *Elective treatment*—it can be postponed easily for the pregnant patient after parturition. However, emergency care should be taken into consideration. The best method of treatment is to eliminate source of pain. Thus for removal of caries, an infected pulp or tooth, surgical procedure should be done under small doses of local anesthetics to minimize use of systemic drugs. Dental procedures are best performed in 2nd trimester for benefit of fetus and optimal comfort of pregnant women.
- *1st trimester*—it is roughly 12–13 weeks. In this first 12 days from conception to implantation known as 'pre-implantation period', exposure to harmful drugs can kill the embryo. From the 13th day, there is period of organogenesis and so the fetus is susceptible to insult and injury resulting in malformation.
- *2nd and 3rd trimester*—after completion of organogenesis, there is considerable growth and development of existing structures like teeth, bones, CNS, endocrine, genitals and immune system. Malformation is less in II and III trimester but drug like streptomycin can still be harmful causing retardation of physical and mental growth,

premature labor or neonatal toxicity. The adverse effect of drugs on the fetus is dependant on the drug and the phase of pregnancy in which the drug is administered.

- *Preventive dental prophylaxis*—preventive dental prophylaxis should be undertaken at the beginning of the 2nd and 3rd trimester.
- *Radiographs*—radiographs are contraindicated in all but emergency situation, when taken lead shielding is mandatory.
- *Reduce chair time*—prolonged chair time must be avoided to prevent supine hypotension.
- *Position*—sitting up position is best for patient since low head position may cause pressure on vena cava and aorta in 2nd trimester
- *Fainting*—in case of fainting, place patient on left side with legs and head elevated. Oxygen and lime juice with glucose could be given and vital sign monitored.
- *Serious complication*—more serious complication such as seizures and active vaginal bleeding or severe cramping require emergency care in hospital.

Guidelines for Prescribed Drugs in Pregnancy

- *Don't use drug unless it is absolute necessary*—use drug in pregnant patient only when it is absolutely necessary.
- *Ruling out possibility of pregnancy*—rule out possibility of pregnancy in every female of reproductive age group and restrict drug usage.
- *Risk and benefit ratio*—prioritize drug usage in the situation and avoid drug usage if the non-usage can do i.e. risk Vs benefit ratio should be calculated.
- *Lower doses*—use lower than usual doses of drug if necessary for short term.

Consideration in Lactation

Almost all drugs are excreted into the breast milk to some extent. An infant normally ingests approximately 1% of the total material dose of a drug. Dental practitioner must remember that drugs should be selected which have short half-life, sustained release formulation should be avoided, drugs should be taken immediately after nursing.

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Emergency Drugs used in Dentistry

5

Introduction

Though rare, life-threatening medical emergencies can and do occur in the dental environment whose incidence is largely unknown. All health care providers, dentists must be prepared to recognize and properly manage patients of medical emergencies in the dental office.

In all cases of emergencies, the best and foremost method of managing medical emergencies is by preventing them from occurring. With this in mind, it is essential to take a comprehensive medical history from any patient who is about to receive dental care.

The following section reviews the more common emergency drugs that the dental team may have to administer in general dental practice.

Emergency Management

First and foremost in emergency management is the ability to effectively provide basic life support, or BLS (includes cardio-pulmonary resuscitation), when appropriate. It is recommended that all dental health care professionals receive regular training in BLS for health care providers, because these skills are maintained only through repetition.

For certain individuals, like those who practice in the remote areas where medical services (routine or emergency) are not readily available, additional training in advanced cardiac life support, pediatric advanced life support, or both may be warranted. Didactic and hands-on training in the prevention, recognition and management of common emergencies also is recommended. Examples of common emergencies include seizures, cardiovascular and respiratory distress, altered consciousness, chest pain and drug related emergencies.

Advanced cardiac life support training involves the following:

- Adjuncts for airway control and ventilation (including intubation)
- Patient monitoring and dysrhythmia recognition.
- Defibrillation and synchronized cardioversion.
- Cardiovascular pharmacology.
- Acid-base balance maintenance.
- Venipuncture.
- Resuscitation of infants, including the newborn.

In addition, all dental offices should maintain at least the basic recommended emergency equipment and drugs. The content and design of these kits should be based upon each practitioner's training and individual requirements. Proprietary emergency drug kits are available, but none of these kits is compatible with the needs of all practitioners. It does recommend that dentists, after considering their specific training and special needs, design their own individualized emergency kits if proprietary kits do not meet their needs.

Types of Emergency Drugs

Drugs that should be promptly available to the dentist can be divided into two categories.

- *Essential emergency drugs*—the first category represents those which may be considered essential. (oxygen, epinephrine, nitroglycerine, salbutamol, antihistamines, aspirin).
- *Additional emergency drugs*—the second category contains drugs which are also very helpful and should be considered as part of the emergency kit. These supplementary drugs. (glucagon, ephedrine, atropine, corticosteroids, morphine/nitrous oxide, nalaxone, lorazepam, midazolam, flumazenil).

Emergency Drug Kit

The dental office emergency kit should be as simple as possible. The emergency drug kit describes in the following

section is a simple organised collection drugs and equipments that has been found to be highly effective in managing those life-threatening situations requiring the administration of drugs. However in most emergency situations, drugs are not necessary for the proper management of the patients. First and foremost in the management of these situations will be the steps of basic life support. In the light of the confusion it seems clear that there is an urgent need to try to rationalise the content of the emergency drugs box and to provide clear, standardised guidance on which emergency drugs they should stock and be able to use.

The following section reviews the more common emergency drugs that the dental team may have to administer in general dental practice.

Adrenaline

- **Indication**—it is the drug of choice for the management of the acute allergic reactions especially in the respiratory and cardiovascular manifestations of allergic reaction.
- **Anaphylactic shock**—an allergic (type 1 hypersensitivity) reaction may be precipitated by any material or drug to which the patient has been sensitized. Life-threatening events include cardiovascular collapse (90%), bronchospasm (30%), angioedema (25%), and pulmonary edema (49%). Severity varies, and onset of anaphylaxis may be delayed for up to 6 hours or be biphasic, reoccurring in 5% of patients after clinical recovery. In addition to adrenaline, high flow oxygen should be administered to all patients in anaphylactic shock. Second-line drugs, used to prevent relapse, are chlorpheniramine and hydrocortisone.
- **Acute asthmatic attack**—adrenaline is also used in acute asthmatic attacks.
- **Cardiac arrest**—adrenaline is used highly advanced cardiac life support protocols.
- **Mode of Action**
 - **Contraction of vascular smooth muscle**—adrenaline is a sympathomimetic amine that activates both alpha and beta adrenoceptors. Contraction of vascular smooth muscle (alpha-mediated). An increased blood pressure helps to maintain cerebral and coronary perfusion.
 - **Increase cardiac output**—increased force and rate of cardiac contraction (Beta1-mediated). This action increases cardiac output, which helps to maintain the blood pressure. However, this may be damaging as it increases myocardial oxygen requirements and may precipitate ischemia.
 - **Relaxation of bronchial muscle**—relaxation of bronchial smooth muscle (Beta2-mediated). Increasing the caliber of the airway in acute anaphylaxis helps to

re-establish airflow restricted by bronchospasm and edema.

- **Inhibition of histamine release**—inhibition of histamine release by mast cells (Beta2-effect). Histamine is an important early mediator, responsible for some of the hemodynamic changes encountered in anaphylactic reactions.
- **Undesirable action**—undesirable action includes its tendency to predispose the heart to dysrhythmia and its relatively short duration of action.
- **Administration**—adrenaline may be given intramuscularly, subcutaneously or intravenously.
- **Dose**—adrenaline is available in two concentrations: 1:1000 (reserved for intramuscular and subcutaneous use) and 1:10 000 (for intravenous administration). It is available in ampoules or pre filled. The following table outlines the recommended volume of 1:1000 adrenaline that should be administered according to age during anaphylactic reactions.

Age	Volume of 1:1000 adrenaline (ml)	Dose (µg)
2	0.2	200
3-4	0.3	300
5	0.4	400
6-12	0.5	500
Adult	0.5-1.0	500-1000

- **Side effects, contraindication and precaution**—supraventricular and ventricular tachydysrhythmia may develop. Should be used with caution in pregnant women because it may decrease placental blood flow and may induce premature labor.

Chlorpheniramine

- **Action**—it is an antihistaminic drug alternative drug is diphenhydramine. Antihistamines are useful in the treatment of the delayed allergic response and in the definitive management of acute allergic reaction. Antihistamines act as a competitive antagonist of histamine. They do not prevent the release of histamine from the cell in response to injury, drugs, or antigens but do prevent access of histamine to its receptor site in the cell and thereby block the response of the effectors cell to the histamine. They are also the potent local anesthetics, diphenhydramine and triphelennamine being particularly potent in this regard. Potential side effect of most antihistamines is a degree of cortical depression which causes sedation. Therefore, Chlorpheniramine is preferred over diphenhydramine as it has less sedative effect.

- *Indication*—delayed allergy, definitive management of acute allergy, as local anesthesia in patients having history of allergy to local anesthetics.
- *Side effects, contraindication, precaution*—side effects include central nervous system depression, decreased blood pressure, it causes thickening of bronchial secretions therefore contraindicated in management of acute asthmatic attacks.
- *Dose*—the drug may be given orally, intramuscularly, subcutaneously or intravenously. For the management of allergic reactions localized to the skin, a dose of 4 mg in adults may be administered orally every 4 to 6 hrly, to a maximum of 24 mg/day.

Oxygen

- *Indications*—it is the most important drug in the entire emergency kit. Oxygen is indicated in all medical emergencies except hyperventilation. Oxygenation prevents hypoxia that may be damaging to vital organs such as the brain and heart.
- *Administration*—ensuring Airway Patency before oxygen is delivered, it is essential to ensure that the airway is patent. Foreign bodies, blood, vomit, and soft tissues may occlude the air space. The airway may be re-established by using the head tilt and chin lift or jaw thrust techniques. Airway adjuncts such as oropharyngeal (Guedel) airways may be used in the unconscious patient to hold the tongue in an anterior position away from the posterior pharyngeal wall. These airways are contraindicated in conscious or semiconscious patients where protective reflexes are active.
- *Oxygenation*—if the patient is breathing spontaneously, a facemask with an attached oxygen supply, running at a flow rate between 4 and 6 /min, may be used to deliver oxygen and help prevent hypoxia. Positive pressure ventilatory support may be provided in respiratory arrest.
- *Dose*—during cardiorespiratory arrest, the chest compression to ventilation ratio for adults 15:2 if one resuscitator is present and 5:1 if two are present.

Nitrates and Nitrites (Glyceryl Trinitrate or Nitroglycerine and Amyl nitrite)

- *Indication*—nitroglycerine and amyl nitrite was the first member of this group to be used as coronary vasodilators more than 100 years ago. These vasodilators primarily used in immediate management of chest pain usually caused by angina or myocardial infarction. Glyceryl trinitrate (GTN) is used for the prophylaxis and relief of angina. Angina, defined as discomfort due to myocardial

ischemia, occurs whenever there is an imbalance between myocardial oxygen supply and demand. Atherosclerotic narrowing of the coronary lumen results in insufficient oxygenated blood being delivered to the myocardium during periods of increased activity.

- *Mode of action*—the principle site of action of GTN is smooth muscle. Within the smooth muscle cell, GTN is converted to nitric oxide which activates the enzyme soluble guanylate cyclase (SGC). Activation of SGC results in increased cyclic guanosine monophosphate (cGMP) production, which leads to relaxation of the smooth muscle cell. As the vascular smooth muscle is relaxed and resting blood pressure is lowered, myocardial oxygen requirement is reduced leads to the relief of angina. GTN improves the myocardial oxygen supply: demand ratio by reducing cardiac workload and increasing the blood supply to the myocardium.
- *Side effect, contraindication and precaution*—the chief disturbances occur due to sudden hypotension. Headaches, postural hypotension, and flushing of face, tachycardia, thready pulse and fainting and complete nitrate syncope might occur. Tolerance is developed rapidly. Methemoglobinemia because of its mild hypotensive action, nitroglycerine is contraindicated in hypotensive patients. Because nitroglycerine is an unstable drug in tablet form, it must be replaced, usually within 3 weeks of its initial use.
- *Administration*—Sublingual tablets (300 micrograms). Uptake is delayed because of the initial time taken for the tablet to dissolve. Spray (400 micrograms metered dose). This is preferable to tablets as the solution is rapidly absorbed, no special storage is required.
- *Dose*—if a patient known to suffer from angina experiences chest pain, they should be placed upright to facilitate respiratory movements. In adults, a dose of 0.3 to 1 mg should produce symptomatic relief within 3 minutes. The effective duration of action is only 20 or 30 minutes; thus, dosing may have to be repeated. Oxygen should also be administered to all patients experiencing acute chest pain. If symptoms are not relieved within 10 minutes, myocardial infarction should be suspected.

Nitrous Oxide

- *Indications*—nitrous oxide may be used for its analgesic properties in the initial management of myocardial infarction (MI). The pain experienced during an MI is often more severe and of longer duration than that in angina. It may lead to further deterioration of cardiac function as it increases sympathetic output, which will elevate the oxygen demands of an already starved myocardium.

- *Mode of action*—many theories of the mechanism of action of general anesthetic agents such as nitrous oxide have been proposed. It is thought that these agents alter, either directly or indirectly, the function of the membrane proteins involved in the conduction of nervous impulses.
- *Administration and dose*—a combination of 50% nitrous oxide and 50% oxygen is used to produce analgesia without loss of consciousness.

Anticonvulsant (Diazepam, Midazolam)

- *Indication*—convulsion may occur in the dental office under several circumstances like overdose or toxicity to the local anesthetics, epileptic seizures, and febrile convulsions. Therefore anticonvulsant drug should be kept in the emergency kit. Seizure disorder is characterized by a stimulation of the central nervous and cardiopulmonary and cardiovascular system, followed by a period of depression of these same systems. as the degree of post-seizure depression is accentuated and its duration is prolonged because of the pharmacological action of the barbiturate. The benzodiazepines, unlike barbiturates, will usually terminate seizure activity without the pronounced depression of the respiratory and cardiovascular system. It is also used in termination of prolonged seizures caused by status epilepticus, local anesthetic toxicity, in hyperventilation and in thyroid storm for sedation.
- *Side effect, contraindication and precaution*—the major side effect of benzodiazepines is respiratory depression and arrest; however, with proper titration during administration, this is unlikely to occur.
- *Dose*—midazolam in 5 mg/ml in 1, 2, 5, and 10 ml vials, diazepam 5 mg/ml in 2 ml ampules.

Methoxamine/Phenylephrine

- *Action*—this one more vasopressor drug is included in the emergency drugs kit because of various shortcomings of adrenaline. Adrenaline is primarily used in the management of the acute allergic reactions and in cases of clinically mild to moderate hypotension. In addition to an increase in blood pressure, adrenaline cause an increase in workload of the heart through its effect on heart rate and cardiac contraction; it also increases the irritability of the myocardium by sensitizing it to dysrhythmias. For this reason, it seems desirable to utilize a vasopressor that will produce to utilize a vasopressor which produce a moderate increase in blood pressure without unduly stimulating the myocardium. Methoxamine and phenylephrine elevates blood pressure through peripheral vasoconstriction.
- *Indication*—in management of hypotension, in which the status of heart is unknown and the intent is to raise the

blood pressure without cardiac stimulation. Possible uses are in syncopal reaction, drug overdose reaction, postseizure states, acute adrenal insufficiency and allergy.

- *Side effect, contraindication, precaution*—vasopressors are contraindicated in patients with high blood pressure or ventricular tachycardia, and such drugs are to be used with extreme caution in patients with hyperthyroidism, bradycardia, partial heart block, myocardial disease, or severe atherosclerosis.
- *Availability*—methoxamine 10 mg/ml or phenylephrine 10 mg/ml.

Antihypoglycemic (50% Dextrose or Glucagon)

Glucagon

- *Indication*—glucagon is a pancreatic hormone that stimulates hepatic glycogenolysis and gluconeogenesis. Glucagon may be given to the unconscious hypoglycemic patient when intravenous access cannot be secured for the administration of glucose. In conscious patients, a sweet drink is sufficient to elevate plasma glucose levels. Oral glucose must not be given to unconscious patients because of the risk of pulmonary aspiration. Hypoglycemia, defined as a blood glucose concentration of less than 2.5 mmol/l, most commonly occurs in insulin-dependent diabetes. Hypoglycemia may result if a patient takes an insulin overdose, patient takes the correct dose but with no food or patient exercises or undergoes a stressful situation.
- *Mode of action*—glucagon increases plasma glucose by stimulating glycogenolysis and gluconeogenesis in the liver. An additional action is inhibition of glycogen synthesis and glucose oxidation. In adipose and hepatic tissues, glucagon causes lipolysis, resulting in the production of fatty acids which further increase gluconeogenesis.
- *Administration and dose*—in adults, 1 mg (0.5 mg in children up to 12 years) may be given intramuscularly into the deltoid or gluteal areas.

Glucose/Dextrose 50%

- *Indications*—glucose may be administered to the conscious or unconscious hypoglycemic patient.
- *Administration*—glucose may be given orally or intravenously. Intravenous administration is the most rapid and effective method of elevating plasma glucose levels.

Salbutamol /Albuterol /Metaproterenol

- *Indication*—asthmatic patients and patients with allergic reactions manifested primarily by respiratory difficulty

will require the use of bronchodilator drugs. These are β_2 adrenergic agonists have specific bronchial smooth muscle relaxing properties with little or no stimulatory effect on cardiovascular or gastrointestinal system. Salbutamol, a potent bronchodilator, is used in the management of acute asthma. Allergic reactions with bronchospasm.

- *Mode of action*—salbutamol is a selective Beta2-agonist which relaxes bronchial smooth muscle.
- *Administration*—to minimize the chance of adverse effects and to produce the most rapid onset of action, salbutamol is administered by inhalation (nebulizer)

Hydrocortisone/Dexamethasone

- *Action*—hydrocortisone, an endogenous adrenal hormone, possesses predominantly glucocorticoids activity and is essential for human survival. Administration of steroids for therapeutic purposes has a negative feedback effect on the hypothalamus and anterior pituitary gland, which eventually leads to atrophy of the adrenal cortex and resultant inability to secrete hydrocortisone in response to stress (acute adrenal insufficiency). Under these circumstances, a patient may collapse due to hypoglycemia and hypotension if a stressful situation, such as dental treatment, is encountered.
- *Indication*—therefore, exogenous hydrocortisone should be administered preoperatively to susceptible patients to restore the normal physiological response to stress. If acute adrenal insufficiency does occur, prompt administration of hydrocortisone and immediate hospitalization is recommended (Dexamethasone and methylprednisolone are contraindicated in acute adrenal insufficiency). Corticosteroids will be administered in the management of an acute allergic reaction, after the acute phase has been brought under control. The primary value of the corticosteroid is in the prevention of recurrent episodes of anaphylaxis.
- *Administration and dose*—hydrocortisone may be administered by the oral, intramuscular or intravenous routes. The dose varies according to the age and situation (25 to 200 mg).
- *Side effect, contraindication, precaution*—except in life threatening emergencies steroid is contraindicated in presence of pre-existing infection, peptic ulcer, and diabetes mellitus.

Atropine

- *Action*—it is a parasympathetic blocking agent, it is recommended for the management of symptomatic bradycardia. By enhancing discharge from the sinoatrial

node, atropine may provoke tachycardia. Atropine will be of benefit in situations in which the patient has an overload of parasympathetic activity on the heart. Extremely fearful patients are likely candidates for this purpose. When stimulated, the vagus nerve acts to decrease SA node activity, thereby slowing the heart rate, when the heart rate become overly slow, cerebral blood flow is decreased and clonical signs and symptoms of cerebral ischemia are noted. By blocking this effect atropine acts to maintain adequate cardiac output and cerebral circulation.

- *Indication*—bradycardia and hemodynamically significant bradydysrhythmia.
- *Side effect, contraindication and precaution*—large dose of atropine may produce clinical signs of overdose, including hot dry skin, headache; blurred nearsightedness; dry mouth and throat; disorientation and hallucinations; atropine is contraindicated in glaucoma or prostrate hypertrophy.
- *Dose*—atropine available in 0.5 mg/ml in 1 ml vial.

Respiratory Stimulant Aromatic Ammonia

- *Action*—aromatic ammonia is a strong respiratory stimulant. It is available in silver gray vaporole, which is crushed and placed under the breathing victim's nose until respiratory stimulation is affected. Aromatic ammonia has a noxious odour and irritates the mucous membrane of the upper respiratory tract, stimulate the respiratory and vasomotor center of the medulla. This action is in turn increases respiration and blood pressure
- *Indication*—vasodepressor syncope, respiratory depression not induced by opioid analgesics.
- *Side effect, contraindication and precaution*—used with caution in patients with chronic obstructive pulmonary disease or asthma. Its irritating effects on the mucous membrane of the upper respiratory tract may precipitate bronchospasm.
- *Dose*—silver gray vaporole containing 0.3 ml of aromatic ammonia.

Drugs for Advanced Cardiovascular Life Support

These drugs should be included only by those doctors who have completed the course in advanced cardiovascular life support. Essential ACLS drugs include epinephrine, oxygen, lidocaine, atropine, dopamine, morphine sulphate and verapamil.

Most of the drugs are already discussed.

Lidocaine

- *Indication*—lidocaine (xylocaine) is considered the primary antidysrhythmic drug in ACLS. It is used

extensively in the management of cardiac dysrhythmias, especially those of ventricular origin that develop after acute myocardial infarction. Lidocaine used in premature ventricular contractions occurring more than six times per minute, sustained ventricular tachycardia and in ventricular fibrillation that is refractory to electrical defibrillation.

- *Side effects*—excessive dose of lidocaine produce myocardial, circulatory, and CNS depression

Dopamine/Dobutamine

- *Action*—dopamine is a chemical precursor of norepinephrine. In large doses it stimulates both α and β adrenergic receptors at lower dose it dilates renal, mesenteric, and cerebral arteries. Dopamine also stimulates the release of norepinephrine; it is indicated for administration in hemodynamically significant hypotension in the absence of hypovolemia. Dobutamine is synthetic sympathomimetic amine that exerts significant inotropic effects by stimulating β_1 and α adrenergic receptors in the myocardium. Its β stimulating action generally overweighs its α stimulating action, usually results in a mild vasodilatation.
- *Indication*—the primary therapeutic indication for dopamine is to treat hemodynamically significant hypotension in the absence of hypovolemia.
- *Side effect*—dopamine may induce or exacerbate supraventricular or ventricular dysrhythmia. It also may imbalance the supply and demand of O_2 to myocardium, inducing or exacerbating myocardial ischemia. Nausea and vomiting frequently are noted with dopamine administration. It is available in 200 mg, 400 mg, 800 mg in 5 ml ampoules.

Morphine Sulphate/Meperidine

- *Indication*—analgesics are used in emergency situations in which acute pain or anxiety is present. Pain or anxiety increases the myocardial workload which increases the O_2 requirement of the myocardium which may worsen the condition. Two such circumstances include acute myocardial infarction and congestive cardiac failure. It is used in acute myocardial infarction, congestive cardiac failure, intense, prolonged pain or anxiety.
- *Side effect*—opioid agonists are potent central nervous and respiratory system depressants. Monitoring of the vital signs is mandatory whenever these drugs are used. Use of opioid agonists are contraindicated in victims of injury and multiple trauma: and should be used with caution in patients with compromised respiratory function.
- *Dose*—morphine is available as 8, 10, and 15 mg/ml and meperidine comes in 50 and 100 mg/ml doses.

Verapamil

- *Action*—it is a calcium channel blocker drug. Verapamil is extremely effective in management of supraventricular tachycardia. It slows conduction through the atrioventricular node, reducing ventricular response to atrial flutter and fibrillation.
- *Indication*—in emergency cardiac care verapamil is primarily used in treatment of paroxysmal supraventricular tachycardia.
- *Side effect and contraindication*—a transient decrease in arterial pressure may be noted because of peripheral vasodilatation. Verapamil is not recommended in ventricular tachycardia; it may induce severe hypotension and predispose the patient to ventricular fibrillation. It is available for injection as 2.5 mg/ml in 2 ml and 4 ml ampoules.

Antidotal Drugs

In order to manage the emergency situations like overdose or toxicity reactions caused by various drugs used primarily for sedation or general anesthesia four categories of antidotal drugs are used it includes the following;

- Opioid antagonists, e.g. Naloxone or nalbuphine.
- Benzodiazepine antagonist, e.g. Flumazenil.
- Antiemergence delirium drug, e.g. Physostigmine,
- Vasodilator, e.g. Procaine.

Naloxone/Nalbuphine

- *Action and indication*—the most significant side effect of opioid agonist is their ability to produce respiratory depression by diminishing the responsiveness of the brain's respiratory center to the arterial carbon dioxide. Naloxone is the only opioid antagonist free of any agonistic properties. It also reverses the other properties of the opioid-like analgesia and sedation. Naloxone may be administered endotracheally in situations where IV access is not available. Improved respiratory function is noted within 2 minutes. Nalbuphine, having opioid agonist-antagonist properties, is used successfully to reverse respiratory depression induced by opioid agonists. And because of its own analgesia-inducing properties it does not entirely remove postsurgical analgesia or sedation.
- *Side effect and contraindication*—naloxone's effect lasts only 30 minutes, respiratory depression may recur if previously given opioid is of long duration. The IM administration of second dose of the naloxone is recommended, its onset is slower but for longer duration than that of IV dose. This minimizes the recurrence of respiratory depression. Naloxone must be administered with extreme care to persons with known or suspected

physical dependence on opioid. Naloxone's complete reversal of agonist effect of opioid may produce severe withdrawal symptoms.

- **Dose**—it is available in 0.4 mg/ml in 1 ml ampoule or 10 ml vial for adults and 0.2 mg/ml in 2 ml ampoules for pediatric administration.

Flumazenil

- **Action**—benzodiazepines are supposed to be most safe drug for anxiety control and sedation. Some side effects like emergence delirium, excessive duration of sedation, and possibly significant respiratory depression do occur. Presence of antagonist for benzodiazepines adds safety to the IV sedation. Flumazenil produce rapid reversal of sedation and to improve the patient's ability to comprehend and obey commands. The duration of antrograde amnesia associate with midazolam was also reduced with use of flumazenil. Flumazenil also decrease the recovery time from midazolam sedation, increase alertness, and provide decrease amnesic effect in geriatric patients. Reversal with flumazenil is not effective following the oral administration of benzodiazepines.
- **Indication**—it is used for reversal of clinical action of parenterally administered benzodiazepines.
- **Side effect**—flumazenil may produce rebound anxiety state in some patients.

Physostigmine

- **Action**—several drugs that are primarily used to induce sedation have the ability to produce emergence delirium also called as anticholinergic syndrome. In this phenomenon the patient appears to lose contact with reality. There may be increase muscular movement, and patient makes unintelligible sounds. Physostigmine a reversible cholinesterase with the ability to cross the blood-brain barrier has become a drug of choice in the management of emergence delirium.
- **Indication**—for reversal of emergence delirium.
- **Side effect, contraindication and precaution**—side effects include increase salivation, possible emesis, and involuntary urination and defecation. If administered rapidly physostigmine can produce bradycardia and hypersalivation. Atropine should always available whenever physostigmine is administered because it is antidote to physostigmine. Physostigmine should not be given in patients with asthma, diabetes, cardiovascular disease, or mechanical obstruction of gastrointestinal or genitourinary tract.

Procaine

- **Action**—whenever there is IM or IV administration of the drug is present, a local anesthetic with significant

vasodilating property is recommended in the emergency kit. Indications for the administration of the procaine are extravascular injection of an irritating chemicals and intra-arterial administration of the drugs. In both cases there is problem of localized tissue irritation and compromised circulation in a local area. Procaine possesses excellent vasodilating properties along with its anesthetic actions, which make it an ideal drug in these situations.

- **Indication**—management of vasospasm and compromised circulation following intra-arterial injection of the drug. And in Management of extravascular injection of irritating drugs or chemicals.
- **Side effect**—allergy to the ester type local anesthetics is not uncommon. Sensitivity test is recommended before administration.

Emergency Equipment

Along with the emergency drugs the emergency equipments are equally important. Personnel who are expected to use these equipments must be well trained in its proper use.

- IV set/cannula/scalp vein set
- Oxygen cylinder
- O₂ delivery system
- AMBU bag
- Syringe and needles
- Tourniquets
- Macgill's forceps
- Suction and suction tips
- Oropharyngeal airways
- Endotracheal tubes
- Laryngoscope

Maintenance of Emergency Kit

- **Freely available**—it should be freely available.
- **Constant place**—the place for keeping the emergency kit should be constant and should not be changed.
- **Sign board**—location of the kit should be highlighted by the placard or sign board.
- **Orientation of working staff**—all the working staff should be oriented towards it.
- **Periodic checking**—it should be periodically checked for the working condition of the equipments, shortfall in the drugs, expiry date of the drugs, and prompt rectification should be made.
- **List of drug**—a list of contents of emergency drug kit drugs along with their expiry date, should be paste on the cover of the kit for the convenience of periodic inspection.
- **Replacement of drugs after used**—whenever the drugs are used they should be immediately replaced.

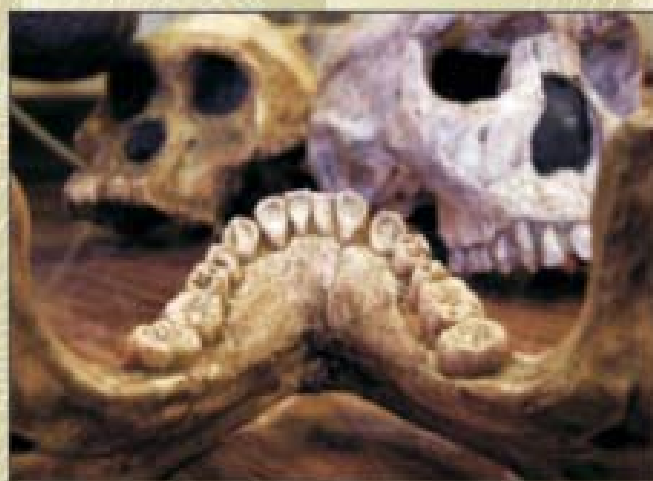
Emergency kit for dental office					
Group	Drug	Use	Concentration	Dose	Route
Anti-allergy	Epinephrine	Anaphylaxis Acute asthma	1:1000	0.3 mg	Subcutaneously
Opioid antagonist	Naloxone	Opioid overdose	0.4 mg/ml	0.4-2 mg	IV or Subcutaneously
Antihistamine	Benadryl	Allergic reaction	10 mg/ml	50 mg	IM, IV
Anticonvulsant	Diazepam	Prolonged seizure	_____	5-10 mg	IM, IV
Oxygen	Oxygen	Respiratory distress	100% inhalation	_____	_____
Vasodilator	Nitroglycerine	Acute angina pectoris	0.4 mg/spray	0.4 mg	Sublingually
Stimulant	Aromatic ammonia	Syncope	Inhalant	0.3 ml	Inhalation
Anti-hypoglycemic agent	Glucose	Hypoglycemia	Few teaspoonfuls	_____	Orally
Analgesic	Morphine	Myocardial infarct	10 mg/ml	10 mg	Orally

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Section **6**

Miscellaneous



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Professional Hazards of Dentistry

Introduction

Medical faculty ensures service to humanity, personal job satisfaction and fulfilment of professional requirements but, it also has certain shortcomings and discrepancies. Dentistry, being an important branch of medical sciences is no exception to this.

Classification

- Professional hazards to dentist from the patients.
- Hazards to the dentist from his working conditions, methodology of practice and materials used.
- Hazards to the dentist from law, i.e. 'consumer protection act (CPA)'.

Professional Hazards to the Dentist from the Patients

Dentistry has an environment, which is at least, dangerous and at worst, lethal. Most often patients suffer from one or the other diseases and due to mass awareness of the importance of these diseases, the problems arise. They are further aggravated by the fact that our profession deals with patient's saliva, blood and mucus which have potential pathogenic microorganisms and are on a constant look out for a portal of entry.

Hepatitis B and AIDS are the most threatening and contagious in almost all cases of infections can be transmitted percutaneously or nonpercutaneously and as dental treatment involves use of small sharp instruments; opportunity always exists for inadvertent percutaneous wounds to operator or the dental staff and thus inevitable spread of infection.

Other infectious microorganisms like viruses, bacteria and fungi can also be transmitted in the dental office and

may either cause a relatively innocuous infection (common cold) or lead to temporary or even long-term debilitation or even death.

Viruses are often shed asymptotically in saliva or other secretions and cause infection to dentist, i.e. herpes simplex virus infecting eyes, hands, fingers. Health care providers can also be a victim of measles, chickenpox, infectious mononucleosis, cytomegalovirus infection, herpes zoster or mumps.

Possibly, dentist gain an infection from patient in contagious stage of syphilis or tuberculous carrier, which is definitely a matter of concern.

Preventive Approach

General

- *Proper medical history*—medical history should be taken thoroughly and should be upto date.
- *Cleaning instrument*—clean the instruments before sterilization, to remove all visible foreign deposits.
- *Sterilization methods*—sterilization methods should be effective against all known pathogens and/or use of autoclave for various instruments. Working surfaces and dental units should be cleaned with chemical disinfectants such as 70% isopropyl alcohol, sodium hypochlorite solution and/or glutaraldehyde.
- *Disposable items*—disposables items like suction tips, impression trays, beakers, needles, towels, masks, and caps should be used.
- *Aspiration and ventilation*—use of high speed aspirators which exhaust externally and proper ventilation will definitely reduce the risk of cross-infection from the aerosols that are formed during certain dental procedures.
- *Management of sharp items*—sharp items like needles, scalpels and local anesthetic cartridges should be placed in a soft container. Collection and incineration of surgical waste should be arranged.

- **Training**—all dental staff should understand the policies for prevention of cross-infection and implement them. Review all the procedures from time to time to ensure that they are being carried out correctly.

Personal protection

- **Gloving**—use of operating gloves and washing hands in between patients with appropriate chemicals are some of the important protective measure.
- **Dental dam**—uses of dental dam provide an excellent barrier from microbial contamination from primary source, i.e. the patient's mouth.
- **Management of cuts and wounds**—cuts and wounds should be covered with waterproof dressings, at all times, even if gloves are worn.
- **Vaccinations**—boosting immune defense of the operator by appropriate vaccinations is compulsory.
- **Face masks and protective eyeglasses**—well fitting face masks and protective eyeglasses should be used during high speed cavity preparations or while using ultrasonic scalers and undertaking surgical procedures.

Professional Hazards to the Dentist from his Working Conditions, Methodology of Practice and Materials Used

Psychological Stress and Fatigue

- **Causes**—psychological climate in a dentist's office is as equally important as is technical expertise. Dentistry has a reputation of being a stressful occupation. Worrying about how others expect you to act, counteracting depression, pressure of keeping time schedules, dealing with anxious and uncooperative patient, law suit threat are all common sources of stress.
- **Burnout syndrome**—we all appear to be prime candidates for the 'burnout syndrome' a syndrome of emotional exhaustion and cynicism frequently seen amongst professionals involved in direct communication with patients.
- **Prevention**—you should cope up with and/or resolve the origin of cause and develop a pattern of healthy living.

Musculoskeletal Complaints

- **Causes**—we all are guilty of adopting bizarre positions while treating our patients and even though such technique cause minimal patient discomfort, we as dentist, are in turn rewarded with muscle fatigue, lower back aches, shoulder problems, etc.
- **Prevention**—reduction of physical stress to the body by adopting a correct posture is important in everyday practice. Low seated, closely supported dentistry with

certain physiotherapeutic exercises provide a new ray of hope.

Cardiovascular Complaints

- **Causes**—it includes varicose veins, high blood pressure and occurs due to erect posture adopted during long standing operating procedures.
- **Prevention**—proper positioning of doctor should be maintained.

Radiation Hazards

- **Cause**—they are responsible for causing changes in blood, skin problems, tumor development, shortening of life span and mutations.
- **Prevention**—use of lead partitions, lead aprons and cubicalization of patients in protective cabins of lead glass. Regular check-up of machines for radiation leakage. Periodic health checks-up of the operator.

Sound Hazards

- **Causes**—air driven, high speed handpieces operating from 3900 to 12,500 Hz and electric engines are main contributing factors towards sound pollution. Hearing loss, acoustic stress and impaired power of concentration first affect the individual who are regularly exposed to frequencies above 3000 Hz.
- **Prevention**—personal evaluation by routine otologic and audiometric examination. Noise attenuation by repair and replacement of defective items, especially high speed drills. Treatment rooms should be made acoustically satisfactory. Personal protection by using ear plugs and muffs which reduce high intensity sounds by 30 to 35 dB.

Light Hazards

- **Causes**—dull light or too much bright light can be hazardous. Ultraviolet radiation used to polymerize sealant and composite restorations can be hazardous and may cause skin cancer and chromosomal changes.
- **Prevention**—protective eye glasses for clinician, auxiliaries and patient with regular ophthalmic consultation is a must.

Hazards due to Materials Used

- **Mercurial hazards**—it can be absorbed through skin and gastrointestinal tract but more commonly, poisoning results from absorption of vapors via lung. It is discussed in detail in chapter of pigmentation. Prevention should be done as follows
 - **Education**—health education regarding hazards due to mercury toxicity.

- *Structural design*—well-ventilated operatory and a proper structural design.
- *Floor covering*—floor covering of nonporous polyvinyl chloride.
- *Avoid room heaters*—avoiding room heaters and sterilization equipments in working areas, to minimize evaporation of mercury.
- *Proper storage*—proper storage in sealed plastic containers.
- *Nontouch technique*—use of nontouch technique during preparation of mercury.
- *Cleaning*—immediate cleaning of mercuric spills and use of water sprays or suction, to prevent release of mercury dust.
- *Periodic health surveillance*—periodic health surveillance by measuring mercury vapor levels.
- *H₂O₂ 30%*—accidental fall of such chemical can cause itching, bleaching and considerable burning of the affected part.
- *Nitrous oxide*—if used in high concentrations during anesthetic administration, it may cause increased absorption and thereby liver and kidney damage with neuralgic disease and congenital abnormalities.
- *Volatile oils*—oil of clove and eucalyptus oil used in dentistry can cause mild burns.
- *TCA and phenol*—their overzealous use can injure both, the patient and the doctor.
- *Root canal sealers*—they have one or more contact allergens and thus cause hypersensitivity and dermatological problems.

Consumer Protection Act (CPA/COPRA)

Surgeons are, at times, the privileged targets of school of law. Intentional damage to the patients and his minors definitely reaps a liability suit to the dentist but in many cases, the innocent surgeons are clutched by these laws thereby damaging the personal and professional ethics.

Introduction

In recent period, the CPA has created a great stir amongst the medical profession on the ground that it would be extremely damaging to the profession and the public service. It was brought into existence in 1986 during which medical service was not included. In April 1995, the national commission, on appeal from Kerala state commission decided that medical service should be covered under CPA.

Consumers section 2(d) of CPA defines consumer is a person who buys goods for a consideration which has been

paid or promised or partly paid and partly promised under any system of different payment. He is the one who hires or awaits of any service or services including any beneficiary. It includes user or beneficiary of goods or service, other than the person who actually buys goods or hires / awaits service where such use is made with the approval of the purchasers.

According to section 2 (o) of the act service means service or any description which is made available to potential users, but does not include the rendering of any service free of charge or under the contract of personal service.

Purpose of Act

CPA 1986 is not a substitute for the existing civil remedies. The redressal machinery set up under the CPA 1986 is an additional facility as stated under section (3) of the act itself. This section says that the act shall be in addition and not in derogation of provision of any other law for the time being in force.

The CPA was brought into existence for the protection of interest of the consumer and for the settlement of consumer disputes with limited time frame and with fewer expenses. This enables consumer to make a complaint to a redressal forum in respect of defective service, if the service has been paid for.

It provides for establishment of consumer councils and other authorities for the settlement of consumer disputes and for matter connected therefore.

The district, state and national commission empowered as '*quasi-judicial bodies*' has been established which looks into complaints of consumers where deficiency of service have come to the notice. These quasi-judicial bodies observe the principle of natural justice and are empowered to give relief of specific nature and award, wherever appropriate, compensation to consumer.

Advantages

- *Free*—administration of justice under the CPA is totally free. Consumer court do not levy court fee in respect of legal proceedings.
- *Speedy justice*—consumer courts are expected to deliver speedy justice.
- *Own lawyer*—you can be your own lawyer before consumer courts, though appointment of lawyer is not prohibited. Consumer court does not encourage appearance of lawyer and extensive long-winded arguments.
- *Procedural simplicity*—procedural simplicity and amicable atmosphere prevailing in consumer courts is more encouraging to an ordinary litigant as compared to lengthy and procedure oriented civil court proceedings.

Machinery

- *District level*—district consumer dispute redressal forum to be chaired by district judge and two other members, one of whom should be a man of good reputation and other should be a lady social worker. At district level, claim for compensation towards damages was fixed to a maximum of one lac at the starting which has been enhanced subsequently to rupees of five lacs in 1993.
- *State commission*—it is for cases where compensation is claimed for more than rupee five lacs but less than twenty lacs. The complaint should be lodged before the state consumer dispute redressal commission which is chaired by high court judge and two other members as selected in case of district redressal forum. In this forum, an appeal against orders passed by district forum can be made.
- *National commission*—it is for cases where compensation claimed for is more than twenty lacs. The complaint has to be lodged before the national body, i.e. national consumer redressal forum. This body is constituted by a judge of Supreme Court selected by Union Government to act as a president of forum with four other members including a lady member. It has been empowered to consider appeals arising from orders passed by any state commission. It is the apex body of redressal forum and has a revisional power.

Procedure

- *Filing of complaint*—complaint is filed by consumer or any voluntary consumer organization registered under Society Registration Act, 1860 or under Company Act, 1956.
- *What complaint means*—complaint means any allegation in writing made by complainant in regard to one or more of the following:
 - That he has suffered loss or damage as a result of any unfaired practices adapted by any doctor.
 - The service mentioned in complaint suffers from deficiency in any respect.
- *Timing of complaint*—as provided under section 24(A) of CPA the complaint has to be filed within two years from the date on which cause of action arises. The complaint has to be filed in any redressal forum subject to its jurisdiction.
- *Content of complaint*—a complaint should contain
 - Name and description and address of complainant.
 - Name and description and address of opposite party.
 - The fact relating to complaint and when, where it arose.
 - Document if any in support of allegation containing the complaint.
- The relief which the complainant is seeking.
- The complaint should be signed by complainant or his authorized agents.
- *Period for giving reply*—after receiving the complaint copy of complainant, it has to be sent to opposite party directing him to give his version case within a period of 30 days which may be extended to 45 days.
- *Chance for appeal*—appeal against order of district forum lies to state forum which in turn lies to national commission.
- *Timing of appeal*—appeal has to be filled within 30 days of verdict. No fees have been prescribed for filling the appeal. Appeal has to be accompanied by certified true copy of order of district forum and reasons for filing appeal should be specified.

Medical Service and CPA

- *Charged service*—in the act itself, there is no mention of medical service but a supreme court judgment by a bench comprising of three judges have held that doctors and hospitals who render service as medical practitioners are accountable for any act of medical negligence and ruled that they can be sued for compensation under the CPA provided the service has not been rendered free. Service rendered to a patient by registered medical practitioner, except where doctor renders service free of charge to every patient or under a contract of personal service by a way of consultation and treatment, both medical and surgical would fall within the ambit of services.
- *Free service will not come under CPA*—services rendered free of charge by RMP attached to a hospital or nursing home or a medical officers employed in a hospital or a nursing home, where such services are rendered free of charge to everybody, would not be the service as defined in the act.
- *NGO service will not come under CPA*—services rendered at non governmental hospital or nursing home where no charge, whatsoever is made from any persons awaiting the service and all patients (rich and poor) are given free service is outside the expression 'service'.
- *Paid NGO service*—services rendered at a non-governmental hospital or nursing homes; where charges are required to be paid by persons, who are in position to pay and rendered free of charge for patients who can not pay fall within ambit of expression 'service', irrespective of the fact that the services are rendered free of charge to persons who are not in position to pay.
- *Free government service*—services rendered at the government hospital health center or dispensary where no charge whatsoever is made from any persons awaiting the service and all patients are given free of

service is outside of preview of the act. The payment of token amount of registration purpose only at hospital or nursing would not alter the position.

- *Paid government service*—service rendered at the government hospital health center or dispensary, where service are rendered on payment of charge and also render free of charge to other persons awaiting such service, would fall of the fact that service is rendered free of charge to person who do not pay for such service.

III Effects of CPA

- *Doctor patient relationship*—it is contention in medical profession in general that the act itself during formulation, did not mean to include the services of a doctor rendered to his patients as doctor patient relationship is something more than consumer trade relationship. This act will totally disturb the doctor patient relationship which is considered to be noble.
- *Extreme laboratory investigation*—it will lead to undesirable tendency in doctors, particularly in general practitioner and new graduates, to be more invasive of their responsibilities towards the patients and refer more number of patients or cases to specialists and to advise extensive laboratory investigations which will make their position comparatively safe; without the improvement of clinical mind and experience; thereby making the treatment of general population to be costlier even up to the extent of being beyond the reach of many.
- *More patient treatment charge*—doctors will develop a tendency to assure himself free from dangers of paying compensation by sticking to different professional indemnity insurance firms which are bound to crop-up to make a good business by taking advantage of this system. As a consequence of all this, patients will be charged more than what they are presently charged.

Introduction

Forensic odontology is the subject concerned with the application of medical and paramedical science knowledge to certain branches of law, both civil and criminal. The medicolegal information obtained from the examination of teeth and jaws falls in the preview of forensic odontology. Upon exposure to physical injury and putrefaction, the human dentition, the enamel of which is the hardest substance in the body outlasts all other tissues. The material used in restoration is also extremely resistant to destruction by chemical and physical elements. The fundamental principles of dental identification are those of *comparison* (when antemortem records of the proposed deceased are available) and *exclusion* (when antemortem records of other persons are available). In the absence of the antemortem records attempt to elicit dental information is made by interrogating the relatives and friends of the deceased, which may frequently prove unreliable.

Forensic odontology may be defined as the application of dental science to the administration of the law and the furtherance of justice.

Scope of Forensic Odontology

- *Record preparation*—the correct handling and examination and the proper preparation and presentation of dental evidence in both civil and criminal legal procedures.
- *Identification*—personal identification, either individually or in context of mass disasters.
- *Age assessment*—to calculate the age of patient.
- *Bite mark investigation*—investigation of criminal cases where bite marks are involved and the interpretation of bite marks.

- *Human abuse*—recognition of domestic, and child abuse.
- *Lip print*—comparison and identification of lip print.
- *Legal aspect*—legal aspect of dental traumatology.

Record Management

- *Content*—the dental record is a legal document of dentist which contains information about subjective and objective finding of the patient. It also includes pathological report, radiographs, and clinical photographs of the patient.
- *Treatment plan*—treatment plan which is given to the patient should be updated in the record. All the letter of reference, letter of consent, insurance and financial statement should be store in the record of the patient.
- *Progress note*—the progress note of the patient should contain information about the restorative and therapeutic procedure which are carried out on to the patient.
- *Record of telephonic conversation*—summaries of the telephonic conversation with the patient, consultant, insurance company representative and legal authorities should be maintained in the record.
- *Signing of record*—record should be signed by the personnel. Any change made in the record should not erased but a line should be crossed on it, so that it is readable. This will help to remove any fraud intention to alter record.
- *Electronic maintenance of dental record*—it is common nowadays that dental record to be maintained electronically. Nowadays some software programs are developed to maintain patient dental information. This is advantageous as it can be easily networked and transferred.
- *Storage of record*—record should be kept minimum of 7 to 10 years. In the case pediatric patient record should be maintain until the patient reaches the age to maturity.

Identification

Forensic odontology is concerned with the identification of both living and deceased person.

Role of Teeth in Establishing the Identity

Dental Comparison

Dental comparison affords a potentially straightforward and simple means of establishing identity. The method of dental identification depends upon:

- *Resistance of dental tissue*—the relative resistance of the mineralized dental tissues and dental restoration to changes resulting from decomposition or harsh environment extremes such as conditions of temperature and violent physical forces.
- *Individual characteristic of dentition*—the unique individual characteristic of the dentition and dental restoration.
- *Antemortem record*—the availability of documentation of the antemortem status of the dentition in the form of dental treatment records and diagnostic radiographs.

Each individual has 32 teeth with 5 surfaces each with their own character of size, shape, position and spacing with the result that no 2 sets of teeth are alike (**Fig. 50-1**). Teeth extracted after death leave a completely different socket from those removed during life. When the tooth is removed or dental work of any sort is carried out the teeth pattern is changed and its record may exist with the dentist. In natural decomposition teeth are practically indestructible. They are not easily destroyed by fire. Being sheltered in the oral cavity they are generally not damaged. Teeth as well as dentures made of acrylic resin are generally resistant to the action of corrosive acids. The identification from data of authenticated teeth depends entirely upon the accuracy and completeness of authenticated records made during life. The record should include:

- *Number of teeth*—the number and situation of teeth present and number and situation of teeth lost.
- *Arrangement*—arrangement, irregularities, erosion, caries, fillings, bridge crown work and dentures.
- *Shape of arch*—exact shape of edentulous arch should be noted down.
- *Identifying features*—some of the common identifying features of teeth pertain to faulty development, faulty alignment, presence of stains, localized wear on certain teeth and missing teeth.
 - *Faulty development*—teeth may be undersized, oversized, notched or present some other irregularity as a result of faulty development and malformation. Hutchinson's teeth constitute a classical example of

malformation of the incisor in congenital syphilis. These changes are most conspicuous in central incisors which are usually small, widely spaced, notched and less broad at the cutting edge than at the gum margin giving them the appearance of tip of screw driver.

- *Faulty alignment*—the defect in the alignment may be in the space between teeth, e.g. widely spaced teeth or overriding teeth. Between the teeth of the upper and lower jaw when there is protrusion of upper incisors resulting in overlap of lateral incisors the bite pattern is known as overbite and the reverse pattern is known as crossbite.
- *Stains*—Pan (betel leaf, tobacco) chewing habit stains the teeth with dark brown or black deposits. Yellowish or dark brown stain on the back of incisor teeth is common in cigarette smokers. Chalky white or yellowish brown areas of discoloration are found in fluorosis. Metal poisoning may cause pigmentation of gums and thereby suggest a cause of death. Copper causes green and mercury and lead a blue black line on the gums. Gum hyperplasia induced by phenytoin may aid in identification and suggest epileptic seizure as a cause of death.
- *Localized wear on certain teeth*—A pipe smoker may have localized wear on teeth either on incisors or at angle of mouth due to position of pipe. Notched incisors from holding thread, pins, nail, between teeth on day to day basis may suggest the occupation of tailor or hairdresser or cobbler.
- *Missing teeth*—The missing tooth may have been lost antemortem or postmortem. Ante mortem loss of teeth due to trauma at or near the time of death if frequently associated with fracture of thin bony plate surrounding the alveolus. In loose tooth which has fallen out it is not so. Extraction or tooth loss in living person is followed by bleeding from its socket which stops in about 24 hrs or sometimes 2 to 3 days when the clot forms in the raw socket. By about 14 days, the clot is obliterated by fibrous tissue and the alveolar rim is smoothened by resorption of bone. By about 5 to 6 months, gradual new bone formation fills the socket but its outline is still visible on X-ray examination. By about 6 months to 1 year remodeling of new bone completely obliterates the socket leaving a slight depression and the socket outline is not visible on X ray examination. In recently recovered remains postmortem tooth loss discloses a clean socket devoid of blood clot. In skeleton in which postmortem loss of teeth is common, the bony rim of alveolus is sharp and feathered.

Procedure in Dental Identification

- *Area covered*—it should include not only the oral cavity of the victim but when applicable, the surrounding scene as well, especially in case of conflagration or injuries when only the remnants of the dental arches may remain scattered and rubble and debris. Dental examination in mass disaster victims often by necessity must be accomplished under accident site and/or temporary mortuary.
- *Recovery of dental structure*—the task of dental identification begins at the site of discovery of the body. When the gross postmortem changes affecting the teeth have occurred, such as charring, disintegration and fragmentation in fires and high impact accident, meticulous care in their recovery and conveyance to the mortuary are of utmost importance. Displaced teeth in decomposed bodies or skeletal remains should be saved, labeled and later secured to the intraoral position using adhesive cement (**Fig. 50-1**). Most open alveoli are the result of postmortem tooth loss. In recently recovered remains postmortem tooth loss disclose a clean socket devoid of blood clots.



Fig. 50-1: Human dentition consist of 32 teeth with 5 surfaces.

- *Instrument*—instruments used for dental examination include explorer, mirror, tissue forceps, heavy duty autopsy scalpel, handle and blades, tissue clamps, irrigating syringe, rubber autopsy gloves, polythene specimen bags, gauze sponges (for tooth cleansing) and a source of illumination (flash light or battery operated head lamps); and dental examination form should be used.
- *Reconstruction and examination*—examination should be performed by two persons thoroughly familiar with dental terminology, one actually performing the examination and one recording the data. The recorder should view the actual teeth in order to record the basic morphological pattern of the restoration or cavities. In

some cases, it is necessary to remove the jaw from the body for more detailed examination and future reference.

- *Transcription of dental records*—the point to be considered while charting are missing teeth, unerupted teeth, supernumerary teeth, restoration, prosthesis, dentures, decayed, broken teeth, mal position, overlapping, crowding and spacing, peculiar shape of teeth.
- *Identification of edentulous bodies*—frequently dentures are present in the mouth of unknown bodies or may be found elsewhere. If the denture can be identified and it can be shown to fit the mouth of the deceased, a reliable identification can be made. The most reliable means of identification of denture is for them to be permanently marked with the name of the patient or some code during manufacture.

Problems in Identification

Condition of material recovered

- It depends upon the circumstances surrounding the death and the care exists in its collection and transport.
- Incineration produces damage to teeth ranging from mild scorching of the surface to severe charring of the enamel and dentine with crumbling of the crown.
- Sustained very high temperature will result in calcinations of the teeth with considerable overall shrinkage.
- Burnt teeth are usually very fragile and suffer separation of the enamel and often gross disintegration of the crowns.
- In high impact accidents such as aircraft and high speed road crashes much mechanical damage can occur and teeth and jaws may fracture and disintegrate.
- Failure to recover significant material may result in failure to identify a body.

Errors in examination—errors can easily be made in the examination and recording of the postmortem dental material.

Inadequate antemortem data—errors in charting teeth treated and insufficient descriptive details about the treatment provided are common. Other difficulty arises when a dentist has retired and destroyed his records.

Technique for Identification in Mass Disaster

These include X-ray, UV light, postmortem serology and DNA profiling.

X-ray

All bodies which are found under suspicious circumstances and which are rendered unrecognizable due to prolonged immersion in water, burning by fire and acid or by any other destructive means such as explosion should

be routinely X-rayed. A dental radiograph when available constitutes one of the most valuable pieces of evidence for identification purpose. Panoramic X-ray technique provides excellent pictorial dental record (**Fig. 50-2**).

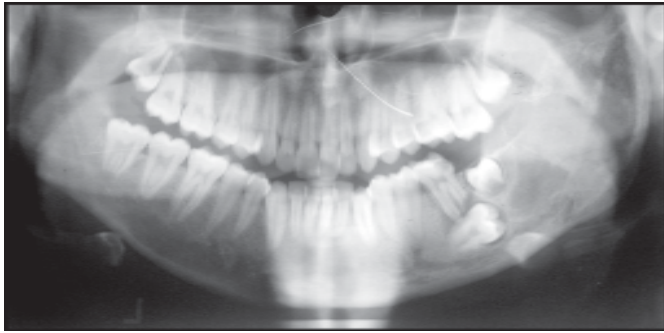


Fig. 50-2: Panoramic radiograph provides excellent record.

The CAPMI (computer-assisted postmortem identification) system compares dental record of victims of mass disaster and enables rapid identification of air crash, flood and explosion victims. Radiography can provide information in relation to age, sex, race, and occupation, diagnosis of certain conditions and identification and cause of death.

Age—Age can be established by radiography of bones and teeth (for root calcification). Calcification of costal cartilage and osteoarthritic changes in large joints and the spine also help.

Sex and race—may be deduced by radiography in some cases.

Occupation—this can sometimes be deduced from X-ray. The whole range of pulmonary occupational diseases such as silicosis, asbestosis may show specific radiographic findings. The radial artery in laborer's using pneumatic drill may show calcification; coal carriers and professional wrestlers are liable to calcified lesions of the ligamentum nuchae. Football players may show calcified hematoma of the thigh muscle.

Identification—it is possible by comparison of postmortem and antemortem X-ray (**Fig. 50-3**).

Cause of death—fracture of bones seen on X-ray may indicate their antemortem origin and these include depressed fracture of skull, fracture of hyoid, fracture dislocation of cervical vertebrae, severe injury to bones by cutting instrument or fracture of several ribs which are incompatible with life. Foreign bodies in the upper respiratory tract provide valuable clue. Evidence of poisoning by heavy metals and signs of diseases such as malignant growth may be apparent.

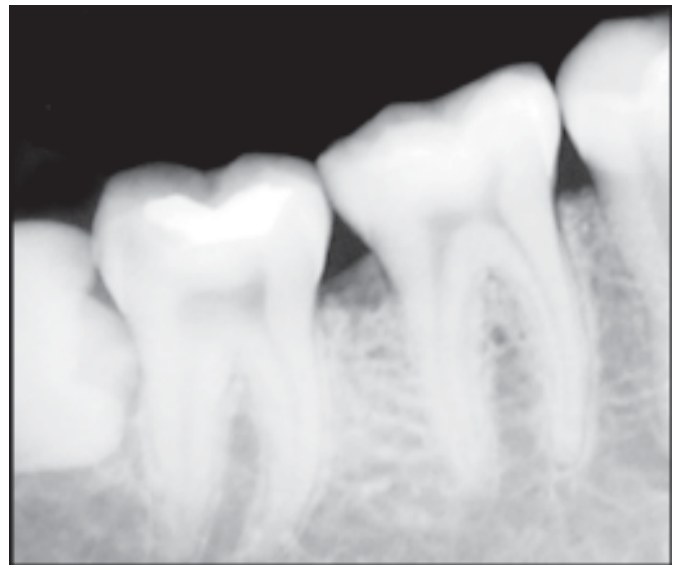


Fig. 50-3: Intraoral periapical radiograph will help to identify the victim on the basis of radiopaque filling.

UV Rays

An ultraviolet lamp can be used to locate and define tattoo marks and scars on burned and decomposed remains, and to segregate bones in cases of mix-up. When examined by UV light washed blood stains are readily seen and seminal stains give a bluish white fluorescence.

Postmortem Serology

A known postmortem grouping of an individual serves to narrow the range of possible identities. Even in putrefied bodies, blood group antigen may be detectable for serological studies. The bone marrow in skeletal remains may still retain serologically detectable antigens.

DNA Profiling

This is useful if suitable tissue (blood, semen stored in bank) is available. If such tissue is available a DNA profiling or autopsy derived tissue should be compared by single probe analysis with that of parent, children, sibling and if be necessary other relatives. This is now used worldwide in aircraft and other major accidents. The techniques which are used for DNA profiling are Restriction fragment length polymorphism (RFLP) and polymerase chain reaction (PCR). RFLP results in splitting source DNA into thousand of fragment. PCR can do evaluation of denture DNA or minute quantity of DNA.

Age Assessment

Chronological age assessment may be an important factor in establishing the identity of the living or deceased person.

It is also important in legal proceedings when specific charge for particular offence may depend on whether the alleged offender is a juvenile.

Visual observation—stage of eruption of the teeth and evidence of changes due to function such as attrition can give an approximate estimate of age.

Radiography—it can provide a great detail, the gross stage of dental development of the dentition.

Histological—it requires preparation of the tissue for detailed microscopic examination, which can determine more accurately the stage of development of the dentition.

Physical and chemical analysis—it is done to determine alterations in ion levels with age have been proposed.

Bite Marks

One of the two major interests of the forensic odontologist is one that has direct relevance to the pathologist, in that it concerns the interpretation of trauma to the body surface. Dental evidence is used to identify the perpetrators of a crime who happened to have left their teeth marks in some substances left at the scene. Generally, it is easier to exclude a certain person than to identify one conclusively from the bite mark.

Definition

A bite mark is a patterned injury produced by teeth on animate or inanimate objects, is caused by small enamel defects on the incisal edge of incisor teeth creating individual characteristics during biting procedures. It can be:

- *Tooth mark*—mark left by a tooth (human or non-human).
- *Arch mark*—mark produced by four or five adjacent teeth in the same arch (**Fig. 50-4**).



Fig. 50-4: Bite marks on inmate subject presented as arch.

Causes

- *Child abuse*—it can be found anywhere on the body, favorite sites being the arms, hands, shoulders, cheeks, buttocks and trunk. Most of the time it is inflicted by the mother.
- *Sexual assaults*—it usually occurs in cases of rape. Most common site of it is breast and nipples but neck, shoulders, thighs, abdomen, pubis and even vulva may be attacked.
- It may also be inflicted on police officers when attempting to arrest resisting offenders.
- *Sports*—bites can occur in sporting events especially football and some forms of wrestling. In this, it can occur anywhere but hands, fingers, nose, forearms, ears and even lips may be target.
- *Self inflicted*—falls onto the face or a fit may cause the tongue and lips to be badly bitten. Some persons deliberately bite themselves, sometime to fabricate injuries for a variety of motives ranging from gain to psychiatric disorders.

Importance of Bite Marks

It can permit precise identification because the alignment of teeth is peculiar to each individual (**Fig. 50-5**). The mark may be on the article of food found at the scene of crime or on human being to realize that a bite on human flesh can have marks not necessarily due to break in the continuity of the skin but due to a small subdermal or thin deep hemorrhage.



Fig. 50-5: Bite mark is represented distorted image of human dentition.

Bite marks are always contaminated by saliva and therefore contain amylase, ptyalin and blood group which can be determined by the cast of suspect of mouth is made and transparency of his bite compared to that of the unknown bite. It should be remembered that bite marks on skin are modified by its elasticity when the teeth are withdrawn.

Method of Preservation of Bite Marks

The doctor should be asked how the bite mark may be best preserved. When the substance is plastic such as butter, cheese, lard, or chocolate, it should be stored in a refrigerator to prevent melting or gradual flowing. It should not be deep frozen as this may cause brittleness and cracking. Fruits should be preserved in Campden solution, a metabisulphite fluid used for fruit bottling. If it is not there then 5% acetic acid in 40% aqueous formaldehyde solution can be used.

Whatever preservation is recommended, the object should be adequately photographed with film plane at right angles to the bite and a scale placed in the focal plane. Death freezes a bite mark but a subdermal hemorrhage in living person disappears within 20 minutes hence it is important to take an immediate photograph of the bite mark.

Classification of Bite Marks

- *Non-human (animals)*
- *Human*
 - *In foodstuffs* (in a part eaten foodstuff abandoned by offenders at the scene of crime).
 - *On non-biological object* (on pencils, pipe stems and detonators).
 - *In human skin*
 - Non-criminal (erotic bite)
 - Criminal—it can be offensive (upon victim by assailant) and defensive (upon assailant by victim).

Bite Marks on Foodstuff

Foodstuff bite marks can be classified into 3 types:

- *Type I*—bites that are found in material such as chocolate, which fracture readily with limited depth of tooth penetration.
- *Type II*—bites formed in foods where teeth obtain a good grip and then bitten piece is removed by fracturing it from main material, e.g. apples.
- *Type III*—bites in which teeth bite right through the bitten material, e.g. cheese. These bit exhibit extensive scrape marks and may give an indication of the relative position of upper and lower incisor teeth in centric occlusion.

Depth of penetration of the teeth—where there is only minimum penetration of teeth into the food, the record is of

biting edges and where there is greater penetration the record is of the labial aspect of the teeth.

Time and temperature—bite mark in foodstuff may produce the exact mesial-distal dimension of the teeth provided the record is taken immediately after the bite was made. Dimensional and color changes are expected to occur in the foodstuffs due to effect of temperature.

Bite Marks in Human Skin

Human tissue has been described as one of the least dependable substances for the registration of bite marks as the bite marks in the tissue is affected by several variables.

Status of the Tissue

Site—type of tissue/condition of skin. For example loose skin or excessive subcutaneous fat commonly demonstrated easy and extensive brushing leading to a poor bite mark definition. Areas of fibrous tissue or high muscle content tend to bruise less easily and thus are more likely to demonstrate a bite mark.

Age—infants and elderly individual tend to bruise more easily than other age groups marking the detection of bite marks more difficult.

Sex—females tend to bruise easier than males. Once produced a bite mark will be evident for a longer period of time on females as compared to males.

In human bite mark can be sexual (**Fig. 50-6**) or assault (**Fig. 50-7**) type.

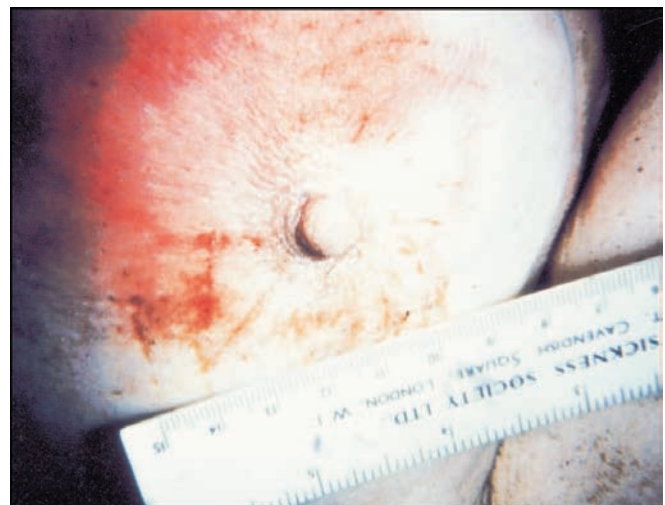


Fig. 50-6: Sexual bite mark.

The Influencing Factor

Bite mark can be influenced by time elapsed between the actual biting and when the impression is made. Depression of the skin as occurs in most bite marks will recover within

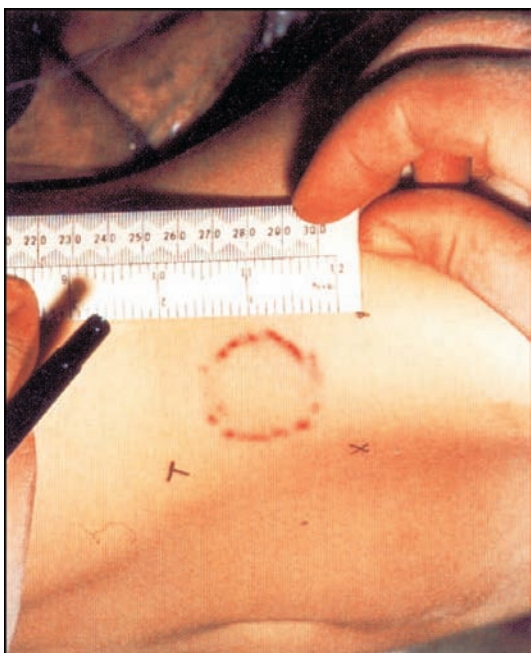


Fig. 50-7: Assault bite mark.

6

10-20 minutes after the bite, although discoloration and swelling may be present 24-72 hours on a living object.

The force exerted—the skin appearance of bite mark may vary from bruising, abrasion, and indentation to actual lacerations.

The number of teeth—in general the more teeth marks present in the bite marks, the better is the likelihood of identification utilizing that bite mark.

The type of teeth—in many cases the bite mark is seen to be comprised of the upper anterior teeth. The investigator should take note of the width of teeth.

The reaction of the surrounding tissue—bite marks made hours or days before the event will show inflammatory changes and signs of healing microscopically.

Types of Bite Mark

- Definite bite mark
- Amorous bite mark
- Moderately aggressive bite mark
- Aggressive bite mark
- Very aggressive bite mark

The Nature of Bite Marks

Though called bite marks, but sometimes it may not be from actual teeth. The lips can transiently mark the skin, if forcibly nipped, especially on children. Suction can produce a crop of punctate hemorrhages, either small petechiae or larger ecchymoses merging into a confluent central bruise.

It is caused by front teeth from canine to canine with a gap at either side representing the separation of upper and lower jaw.

A human bite is near circular or a shallow oval. A deep parabolic arch or 'U-shaped' can only be animal in origin. The teeth may cause clear, separate marks or they may run into each other to form a continuous or intermittently broken line. As the time progresses original teeth marks spread out and blur. Teeth marks may be abrasion, bruises or laceration or a combination of any two of three. The clarity of bite marks depends on a number of factors:

- If the contour of the part bitten is irregular or markedly curved, then only part of dental arch is in contact with the tissues.
- If the bite is forcible, then extensive subcutaneous bruising may spread and blur the outline.
- If the bite is inflicted many days before, then healing of abrasion and lacerations and absorption of bruising will leave progressively less detail.
- When the teeth are forcibly applied the typical appearance is of two 'bows' with their concavities facing each other and a gap at each end.
- *Love bites*—they are caused by firm application of the lips, which form an airtight seal against the skin, and then sucking action reduces the air pressure over the center. This causes shower petechial hemorrhage to appear from rupture of small venules in the superficial layer of the subcutaneous tissue. If forcible the petechiae are confluent and a frank bruise or even hematoma develops.

Investigations of a Bite Mark

Firstly the bite mark should be carefully and fully photographed. The photograph should be taken from different angles, but especially from directly perpendicular viewpoint, with the plane of film at right angle to that of the lesions. An accurate scale should always be held near the lesion, as close as possible. The lesion should almost fill the camera frame in some shots to capture as much detail as possible. When photography is completed, swabs of the bite should be taken to try to recover saliva. Plain cotton-wool swab are gently rubbed onto the bite. They should be then deep frozen unless send straight to the serology laboratory.

After this, impression of the bite can be taken (Figs 50-8 and 50-9). It is done by laying a plastic substance over the bite mark, which then hardens, so as to produce negative cast of the lesion (Fig. 50-10). It is usually made with a rubber- or silicone-based medium containing catalytic hardeners. Less satisfactory substances are water based pastes, such as plaster of Paris, which are put on wet and allowed to dry before removal. But they have a disadvantage



Fig. 50-8: Sample bite taken on apple

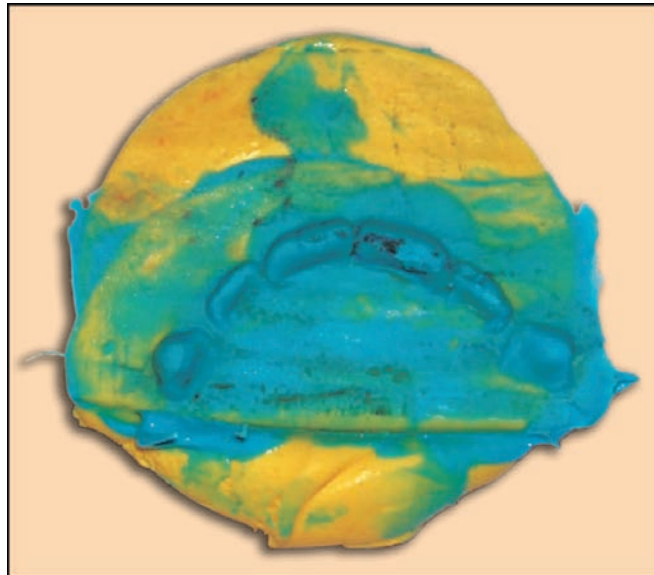


Fig. 50-9: Impression is taken of the bite and cast is made.



Fig. 50-10: Study case is made.

of potential damage to the actual bite. After autopsy, it is also possible for the whole area of the skin carrying the bite to be removed and preserved in formalin for future examination.

Matching the Bite Mark with the Suspect Dentition (Figs 50-11 to 50-14)

The teeth of those who are either suspected by the police or who had access to the victims should be examined. In most jurisdictions, it is vital that fully informed consent should be obtained from the persons beforehand.

Any refusal must be a bar to any further action. When children are concerned, usually in the setting of child abuse, the consent of fully informed parents or guardian must be



Fig. 50-11: Bite mark of suspect is taken.

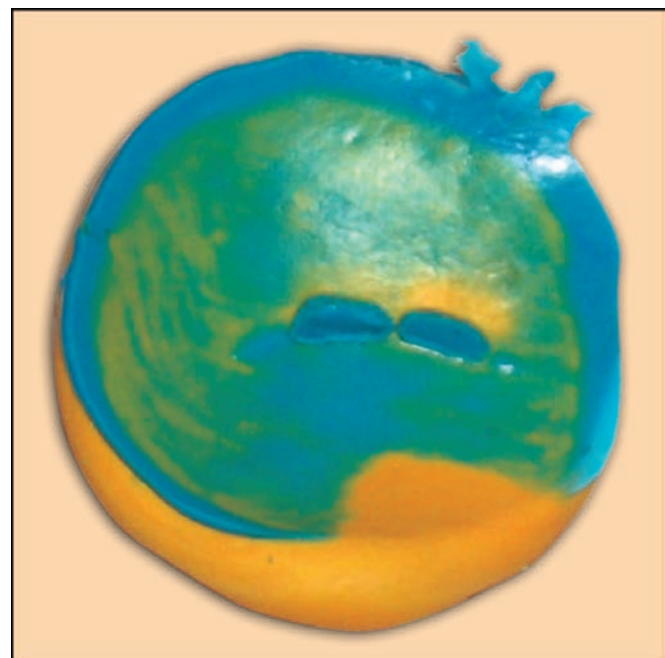


Fig. 50-12: Positive replica of bite mark is made.



Fig. 50-13: Cast of suspect is made.



Fig. 50-14: Cast is matched with previous mark.

obtained. Then dentition is examined for number of teeth, missing teeth, complete or partial denture, occlusion, broken teeth, irregular teeth and abnormalities of the teeth. Photographs of dentition of the patient can be taken.

After that impression of bite of suspect should be taken. Tracing can be made from the positive cast of a bite impression, inking the cutting edge of front teeth and transferring these to transparent sheets, which can then be laid over the photograph to determine correspondence.

Classification of Skin Wounds

Abrasion—it is superficial type of injury only skin part is involved, i.e. both epidermis and dermis. It heals without the scar formation. It is of following types:

- **Graze**—abrasion caused by friction between skin and hard surface.
- **Scratch**—it is caused by pointed ends of nails, pins, needles, etc. It is localized linear abrasion.
- **Imprint abrasion or pattern abrasion**—it gives the pattern of object on the skin. For example teeth bite.
- **Bruise or contusion**—subdermal-extravasation of blood from rupture of subcutaneous vessels and capillaries of skin and organs.
- **Laceration**—wound must be produced with a blunt object that penetrates the skin to subcutaneous tissue.
- **Incised wound**—must be caused by sharp edged object like knife. In incised wound length is more as compared to width and depth.
- **Stab wound**—it is caused by pointed end of sharp edged or blunt object. Depth is more as compared to width and length.

Contusion and abrasion are described in two dimensions, i.e. length and width.

Laceration, incised wound and stab wound described in three dimensions.

Medicolegal Aspects of Dentistry

The Dentist Act 1948

The act was passed to regulate the profession of dentistry in general and to constitute the Dental Council of India and for a register called 'Indian Dentist Register'. It also provides for the constitution and composition of the State Dental Council and for State Register of Dentists.

Duties of Dental Council of India

- To maintain the dental register.
- To regulate dental education.
- To recognize the foreign dental qualification.
- Appeal against disciplinary action.
- To give warning notices to dentist for his/her misconducts.

Duties of State Dental Council

- To maintain dentist register.
- To take disciplinary control.
- To give warning notice to dentist for his/her misconduct.

Hippocrates Oath: The Declaration of Geneva

- I solemnly pledge myself to consecrate my life to the service of humanity.
- I will give my teacher, the respect and gratitude which are their due.

- I will practice my profession with conscience and dignity.
- I will respect the secrets which are confided in me.
- The health of my patient will be my first consideration.
- I will maintain by all the means in my power, the honors and the noble tradition of the medical profession.
- My colleagues will be my brothers.
- I will not permit consideration of religion, nationality, race, party, politics or social standings to intervene between duties to my patient.
- I will maintain utmost respect for human life from the time of conception, even under threat; I will not use my medical knowledge contrary to laws of humanity.

Rights and Privileges Enjoyed by Registered Dental Practitioner

- Right to choose the patients.
- Right to use title and description of the qualification which is actually possessed.
- Right for appointment in public and local hospital.
- Right to prescribe and/ or dispense medicine to his patients.
- Right to realize fee and other expenses.
- Right to issue medical certificates and medicolegal reports regarding dental problems.
- Right to evidence in a court of law as an expert witness.
- Right to exemption from serving as a juror at an inquest.

Duties of Dental Practitioners

- Duty to exercise a reasonable degree of skill and knowledge.
- Duties with regard to attendance and examination.
- Duty to furnish proper and suitable medicines.
- Duty to give instructions.
- Duty to control and warn.
- Duty towards children and adults incapable of taking care of themselves.
- Duty to inform patient of risks.
- Duty to notify certain diseases.
- Duties with regards to operation.
- Duties under Geneva conventions.
- Duties with regard to consultation.
- Duty in connection with X-ray examination.
- Professional secrecy.

Professional Misconducts

- Adultery.
- Improper conduct or association with patient or member of patient's family.
- Conviction by court of law.
- Issuing false certificates.

- Withholding from health authorities information of notifiable disease.
- Performing or enabling an unqualified person to perform an abortion or any illegal operation for which there is no indication.
- Violating the provisions of the Drug Act.
- Selling scheduled poisons to the public under cover of his qualifications.
- Using of touts and agents for procuring patients.
- Disclosing the secret of a patient.
- Association with manufacturing firms.
- Advertising.
- Professional association with bodies or societies of unqualified persons formed for the purpose of turning unqualified practitioner.
- Running an open shop for sales or medicines for dispensing prescription of other doctor or for sale of medical and surgical appliances.
- Refusal to give professional service on religious grounds.
- Drunk or disorderly so as to interfere with proper skilled practice or medicine.

Precautions Against Negligence

- Obtain informed consent of the patient.
- Establish good rapport with the patient.
- Keep full, accurate and legible medical records.
- Employ ordinary skill and care at all time.
- Confirm diagnosis by laboratory tests.
- Take skiagrams in bone or joint injuries or when diagnosis is doubtful.
- Immunization should be done whenever necessary particularly for tetanus.
- Sensitivity tests should be carried out before injecting preparations which are likely to cause anaphylactic shock.
- Seek consultation where appropriate.
- In suspected cases of cancer all laboratory investigations should be done without delay to establish early diagnosis.
- Do not criticize or condemn the professional ability of another doctor especially in the presence of the patients.
- Do not make statements admitting fault on your part.
- Avoid firm overconfident prognosis and promising too much to patients.
- Never guarantee a cure.
- Inform the patient of any intended absence from practice or recommended or make avail of a qualified substitute.
- Transfer the patients if facilities are inadequate to handle his problems.
- The drug should be identified before being injected.
- Obtain consent for an operation or giving anesthesia.
- No experimental procedures should be adopted without the consent of the patient.

- No procedures should be undertaken beyond one's skill.
- Keep yourself informed of technical advances and use standard procedures of treatment.
- Frequently check the condition of equipments and use safety installations.
- Proper instructions should be given to patient and proper postoperative care should be given.
- The patient must not be abandoned.
- No female patient is examined unless a third person is present.
- Do not order a prescription over the telephone.
- Anesthesia should be given by qualified medical professional only.
- Do exercise care in the selection of assistants and allotting duties to them.

Penal Provisions Applicable to Medical Practice

- *S.176 IPC*—omission to give notice or information to public servant by person legally bound to give it.
- *S. 177 IPC*—furnishing false information.
- *S.191 IPC*—giving false evidence.
- *S.192 IPC*—fabricating false evidence.
- *S.193 IPC*—punishment for false evidence.
- *S.197 IPC*—issuing or signing false certificate.
- *S.201 IPC*—causing disappearance of evidence of offence, or giving false information to screen offender.
- *S.203 IPC*—giving false information respecting an offence committed.
- *S.204 IPC*—destruction of document (or electronic records) to prevent its production as evidence.
- *S.39 cr. P.C*—every person aware of the commission of

the intention of any other persons to commit any offence punishable under IPC, shall forth give information to the nearest magistrate or police effective of such commission or intention.

- *304 (A) IPC*—causing death of the patient due to rash or negligent act.
- *320 IPC*—grievous hurt—the following kind of hurt only designated as 'grievous' hurt.
 - *First*—emasculation
 - *Second*—permanent privation (loss) of the sight of either eye.
 - *Third*—permanent privation of hearing of either ears.
 - *Fourth*—privation of any member or joint.
 - *Fifth*—destruction or permanent impairing of the powers of any member or joints.
 - *Sixth*—permanent disfigurement of head or face.
 - *Seventh*—fracture or dislocation of bone or *tooth*.
 - *Eighth*—any hurt which endangers the life or which causes sufferer to be during space of 20 days in severe bodily pain, or unable to follow his ordinary pursuits.

Suggested Reading

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3. Standish SM, Stimson PG. Forensic Dentistry Legal Obligation and methods of identification for the practitioner: *Dent Clin of North Am* 1977;21:1-196.
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Introduction

As there is changing era with more advance diagnostic technology coming up, nomenclature of some of the diseases should be revived and changed. In dentistry, there are many disease names which are nowadays either discarded or had been given new names. But, as it is being said difficulty lies not in new ideas but escaping old one, we many times not accept the newer terms and stick to older ones. This chapter will give emphasis on some of older terminologies and why it should not be used.

Some of the diseases in this chapter are described in detail. The reason for this is that in many parts of the world these terminologies are still used. But as a practice, we should start using new terminologies to avoid confusion in mind of budding dental professionals.

Bowen's Disease

It is localized '*intraepidermoid carcinoma*' that may progress to invasive carcinoma over many years, which is characterized by progressive scaly or crusted plaque-like lesion.

Etiology

- *Sun exposure*—this is thought to be causative factor for the Bowen's disease.
- *Arsenic ingestion*—accidental ingestion of arsenic can lead to this disease.

Clinical Features

- *Age and sex distribution*—it is common in older age group with more predilections for males.
- *Location*—it occurs on male and female genital mucosa and in oral mucosa as erythroplakia, leukoplakia or erythematous lesion.

- *Appearance*—it appears as slowly enlarging erythematous patches.
- *Skin*—there is a red and slightly scaly area on the skin, which eventually enlarges and turns into white or yellowish lesion.
- *Signs*—when these scales are removed, it produces a granular surface without bleeding.

Why Discarded

Bowen's disease nowadays synonymous with carcinoma in situ. Histologically, Bowen's disease is characterized by disorganized growth, presence of large hyperchromatic nuclei and multinucleated cells which are the same features that are seen in carcinoma in situ.

Leontiasis Ossea

The word '*leontiasis*' has been used to describe the leonine appearance of some patients with facial leprosy. Virchow added the word '*ossea*' to describe leonine appearance in bilateral bone disease of the face and thus, the term leontiasis ossea came into being.

Etiology

- *Bone disease*—most cases of leonine face are result of bone disease from two conditions i.e. *fibrous dysplasia* and *Paget's disease*.
- *Metaphysial dysplasia*—bilateral and symmetrical involvement of the face and jaws may be seen in some cases of *metaphysial dysplasia* and in *diphysial dysplasia*.
- *Periostitis of jaw bone*—some cases of leonine ossea are due to *periostitis* of the jaw bones or it may be due to *osteitis fibrosa*.

Clinical Types

- *Early childhood type*—starts early in life and quickly extends until skeletal growth ends, when the lesion ceases to grow but does not regress.
- *Young adult type*—second group starts in early years and while it progresses slowly or quickly, it does not cease with the end of somatic growth but continues its activity and may produce gross and terrible consequences.
- *Adult type*—the third type starts in early adulthood or even near the middle life and progresses slowly and inexorably.

Clinical Features

- *Appearance*—there is bilateral enlargement of the facial bones, sometimes producing the leonine appearance.
- *Site*—a classical case is one in which there is enlargement of maxilla, mandible, and malar bones and in some cases, changes in the frontal as well as ethmoid, sphenoid and temporal bones. In some cases, there is partial deformity of the face, either unilateral or bilateral, but without leonine appearance.
- *Orbit*—the orbital cavity is reduced in size and the eye is protruded. Blindness follows compression of the optic nerve or the eyeball and difficulty in breathing is the result of narrowing of the nasal passage.

Radiographic Features

- *Radiodensity*—the affected bones vary in size and density.
- *Appearance*—there may be symmetrical thickening and increased density of the bones without any leonine faces.
- *Mandibular lesions*—there is tendency of mandible to present a characteristic appearance. The inferior margins of the bone, deep to bicuspid and molar region projects downward, perhaps very markedly, while the incisor area is either unchanged or only slightly deeper than normal.
- *W appearance*—the effect of altered configuration of the mandible is that it represents a rough caricature of the letter W.
- *Cumulus cloud*—the surface of the bony projection from the inferior aspect of the mandible may present irregular appearance resembling the cumulus cloud.
- *Nose*—the nasal bone is often thickened and nasal septum may or may not be involved.
- *Frontal bone*—in some cases there is marked prominence of frontal bone over the orbits.
- *Face*—there is gross deformity of the face, with large mass of bone projecting from the mandible on one side and another, from the opposite paranasal region.
- *Skull*—the base of the skull tends to be either granular in structure or homogeneous and structureless, but very dense.

- *Marrow spaces*—in some cases the trabeculae are thickened and dense, with narrowing of the marrow spaces.

Why Discarded

Nowadays this term is never used as it requires a lot of imagination to see lion in it.

Previous or Former Cyst

Nowadays some of the names which are used to describe earlier are not in use or they are no longer suitable. It includes primordial cyst which is seen to be odontogenic keratocyst, globulomaxillary cyst which nowadays seems to be odontogenic origin as entrapment theory is not accepted. Globulomaxillary cyst is said to be radicular, lateral periodontal cyst or OKC. Other cysts which now said to be not in existence are median mandibular cyst and median palatal cyst which are variants of nasopalatine duct cyst.

Primordial Cyst

It is one of the less common types of odontogenic cysts.

Origin

- *Stellate reticulum*—it originates when cystic changes take place in the stellate reticulum of the tooth germ before any calcified enamel or dentin has been formed. So, it is found in place of a tooth rather than directly associated with it.
- *Supernumerary tooth*—it may originate from the enamel organ of supernumerary tooth or from the remnants of dental lamina.

Clinical Features

- *Occurrence*—less common and account for only 5% to 6% of cysts of the odontogenic variety.
- *Age and sex*—it is found in children and young adult between 10 and 30 years of age, although it may persist in older age group and occurs with equal frequency in both the sexes.
- *Site*—it can arise in any portion of the jaw, but most often seen in the ascending ramus of the mandible and in third molar area. It is occasionally associated with an over-retained erupted deciduous tooth.
- *Symptoms*—it has a tendency to painlessly enlarge and slowly replace large portions of cancellous bone before expansion of the cortical plate by way of which it reveals its presence. Pain which is associated with a large cyst

is caused by infection that may follow the perforation of the expanded cortical plate.

- **Aspiration**—when aspirated, they yield a thick granular yellowish material.

Radiographic Features

- **Radiodensity**—cyst-like radiolucency that is well defined and have hyperostotic borders.
- **Internal structure**—it may be unilocular or have a scalloped outline that gives it a multilocular appearance (**Fig. 51-1**).
- **Teeth**—it produces deflection of adjacent tooth root, but seldom cause any root resorption. The involved tooth is missing because of failure of the tooth to develop.
- Those in maxilla are smaller than their mandibular counterparts.

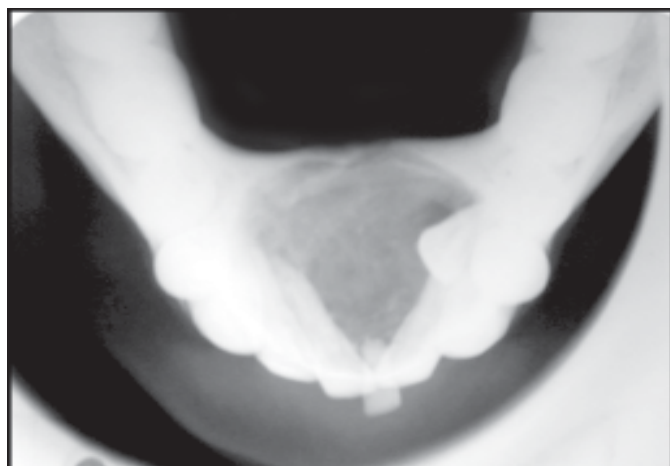


Fig. 51-1: Primordial cyst showing radiolucency in lower anterior region with missing central incisor.

Why Discarded

Nowadays this term is not used and this cyst is thought to be odontogenic keratocyst.

Globulomaxillary Cyst

It is also called '*intra-alveolar cyst*'. It occurs in globulomaxillary area. There is evidence that the cyst acutely forms in the bone suture between the premaxilla and maxilla, the incisive suture, so that location may be different. Due to this Ferenczy has suggested the term '*premaxilla-maxillary cyst*'.

Pathogenesis

- **Fissural cyst**—it was considered to be an inclusion or developmental cyst that arises from entrapped non-odontogenic epithelium in globulomaxillary suture

which occurs at the junction of globular portion of the medial nasal process and maxillary process. Previously existence of globulomaxillary cyst was disputed by Christ who said that development of anterior maxilla occurs by merging of growth center and not by fusion of facial processes so no entrapment occurs because it cannot exist. Recent embryonic studies have demonstrated that Christ's view of facial development is incorrect. Fusion of facial process does occur and epithelium is entrapped in areas that will later lie between the maxillary lateral incisors and canines. In conclusion, recent evidence favors the re-introduction of the globulomaxillary cyst as a specific pathologic entity with varying histologic features.

- **Odontogenic origin**—recent theory suggests most of cyst originating in globulomaxillary area are of odontogenic origin. This is reason nowadays, it is assumed as variant of lateral periodontal cyst, radicular cyst or OKC.

Clinical Features

- **Location**—it is present between maxillary lateral incisor and canine.
- **Symptoms**—it is asymptomatic and is discovered during routine radiographical examination. If cyst becomes infected, patient may complain of local discomfort or pain in that area.
- **Teeth**—as it enlarges, it expands the buccal cortical plate between maxillary lateral incisors and canines. This will diverge the roots of two teeth and their crown resulting in moving contact point incisally. Adjacent teeth are usually vital.
- **Signs**—mucosa over the expanding cortex remains normal in color. If cortical plate is eroded then fluctuant swelling develops. Palpation will produce crepitus. If it becomes secondarily infected, the expansion will mimic lateral periodontal abscess. Aspiration of the swelling is productive of typical amber colored cystic fluid.

Radiographic Features

- **Pear-shaped radiolucency**—it appears as pear-shaped (**Fig. 51-2**) or tear-shaped radiolucency between roots of maxillary lateral incisors and canines. Small end of the pear is directed toward the crest of alveolar ridge. The upper border may invaginate the floor of the nasal fossa or the antrum.
- **Size**—is variable and may reach the maximum level of diameter of 3-4 cm.
- **Teeth**—it may cause divergence of the roots adjacent teeth (**Fig. 51-3**). Displacement of the teeth is common. In some cases root resorption is also seen. The rotation and separation of the lateral incisor and canine roots will usually be apparent on the radiograph.

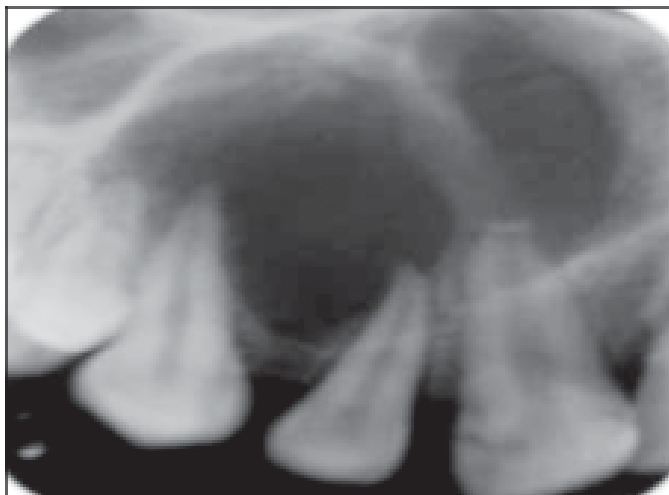


Fig. 51-2: Pear-shaped radiolucency of globulomaxillary cyst seen between lateral incisor and canine. Note there is also root resorption of teeth.

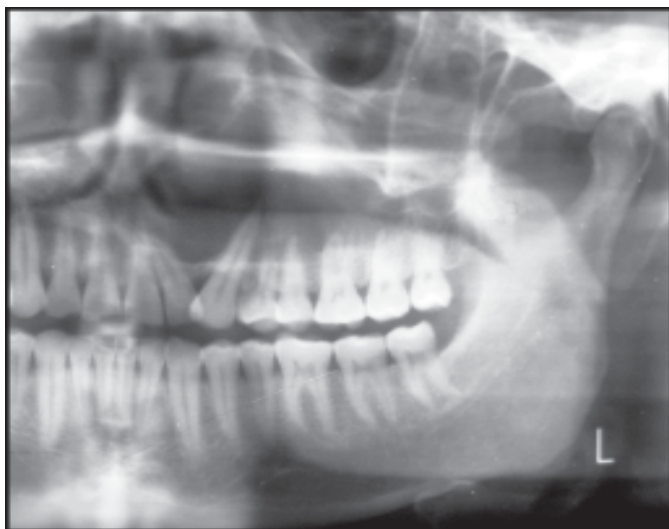


Fig. 51-3: Crop OPG showing radiolucency in lateral and canine region causing the divergence of teeth (Courtesy Dr Ashok L).

- *Lamina dura*—lamina dura of the adjacent teeth are usually intact.

Differential Diagnosis

- *Lateral periodontal cyst*—radiographically, it appears as a dome-shaped radiolucency more commonly seen in mandibular lateral incisor and first premolar region occurring in an older age group.
- *Lateral dentigerous cyst*—it is commonly associated with impacted teeth. The radiolucency is associated with the crown (attached to the neck of the tooth).
- *Primordial cyst*—it is more common in mandibular posterior region.
- *Giant cell granuloma*—it is more common in anterior region. Usually, it appears as a mandibular multilocular radiolucency.

- *Traumatic bone cyst*—it appears as round shaped with moderately defined outline. Needle aspiration is non-productive.
- *Adenomatoid odontogenic tumor*—radiolucency is associated with unerupted teeth. In mature stage, it appears as radiolucent with radiopaque foci.
- *Surgical defect*—patients give history of surgery.
- *Odontogenic myxoma*—in myxoma, mandible is more commonly affected. It shows a typical honeycomb pattern.
- *Anatomical variation*—in some cases, prominent incisive fossa can get confused with globulomaxillary cyst. But location of incisive fossa is in anterior region between central incisors.

Why Discarded and Updated Term

As the study by embryologist shows that there is no process or bony primordia are separated by epithelium. So there is no chance of entrapment of epithelium in this location so use of term globulomaxillary cyst is discarded. Nowadays any cyst in that location will be either radicular cyst from nonvital lateral incisor, or odontogenic keratocyst or adenomatoid odontogenic cyst.

Median Mandibular Cyst

The median mandibular developmental cyst is an extremely rare lesion occurring in the midline of the mandible. It is of questionable existence.

Origin

- *Fissural cyst*—some authors consider it as a true developmental cyst originating from proliferation of epithelial remnants entrapped in the median mandibular fissure during fusion of bilateral mandibular arches. But, as mandible is single bilobed proliferation of mesenchyma with central isthmus, there is no fusion of epithelium line process occurs, so entrapment of epithelium will not occur. So this theory is not possible for the development of median mandibular cyst.
- *Odontogenic origin*—nowadays, it appears that this entity, is a variant of lateral periodontal cyst or odontogenic keratocyst.

Clinical Features

- *Site*—it has got predilection for the inferior part of the mandible, in the central incisor region.
- *Symptoms*—most are clinically asymptomatic and are discovered only during routine radiographic examination.

- **Signs**—they seldom produce obvious expansion of the cortical plate of bone.
- **Teeth**—associated teeth react normally to pulp vitality test.

Radiographic Features

- **Appearance**—it is unilocular, well-circumscribed radiolucency, although it appears multilocular.
- **Shape**—the image is well defined, round or ovoid radiolucency that may be regular or irregular in shape.
- **Lamina dura**—the lamina dura around the lower incisor teeth is intact.
- **Teeth**—as it expands, it diverges the roots of the mandibular incisors.

Why Discarded

This cyst is discarded as there is no possibility to have epithelial entrapment during embryonic fusion in mandible.

Median Palatine Cyst

It is rare fissural cyst that developed due to entrapment of epithelium in line of fusion of lateral palatal shelves of maxilla. It is very difficult to distinguish from the nasopalatine cyst. Nowadays it is known as posterior extension of incisive canal cyst.

Clinical Features

- **Site**—it is very rare and develops in the midline of the hard palate posterior to pre-maxilla.
- **Symptoms**—patient notices the swelling in the midline of palate posterior to palatine papilla. It is usually asymptomatic but some patients may complain of pain.
- **Signs**—it is fluctuant and non-tender. Overlying mucosa is normal. Corticated plate may be perforated as the cyst grows. Expansion is rare in this cyst.
- **Teeth**—maxillary teeth are vital and aspiration produces amber colored fluid.
- If floor of nasal fossa is eroded, cyst may be superiorly displaced.

Radiographic Features

- **Site**—radiolucent lesion is behind the incisive canal in premolarmolar area.
- **Margins**—well-defined borders which are hyperostotic.
- **Appearance**—nasal septum image crosses the septum and appears on occlusal radiographs.
- **Teeth**—in some cases, divergence of teeth may occur.

Differential Diagnosis

- **Radicular cyst**—vitality test and periapical radiographs demonstrate intact lamina dura and normal periodontal ligament space in median palatine cyst.
- **Palatal space abscess**—soft, fluctuant swelling and yield pus on aspiration and non-vital teeth found adjacent to it which give rise to infection.
- **Incisive canal cyst**—it occurs in canal above palatine papillae while midpalatine cyst occurs in midline of palate posterior to palate.
- **Retention phenomenon**—seen laterally and not in midline. Aspiration would not yield amber colored fluid but viscous clear sticky liquid.
- **Malignant and benign tumors of salivary gland**—laterally and not in midline.

Why Discarded

This cyst is nowadays thought to be posterior extension of nasopalatine cyst.

Adenomatoid Odontogenic Tumor

It is also called 'Adenoameloblastoma' or 'ameloblastic adenomatoid tumor'. It is tumor of odontogenic epithelium with a duct-like structure and varying degrees of inductive changes in the connective tissue. The tumor may be partly cystic and in some cases the solid lesion may be present only as masses in the wall of a large cyst. A well-circumscribed lesion derived from odontogenic epithelium that usually occurs around the crowns of unerupted anterior teeth of young patients and consists of epithelium in swirls and ductal patterns interspersed with spherical calcifications.

Origin

- **Dental epithelium**—the close resemblance of the columnar cells to ameloblasts and the frequent association of the tumor with unerupted teeth indicate its origin from dental epithelium.

Types

- Peripheral adenomatoid odontogenic tumor
- Central adenomatoid odontogenic tumor
 - **Follicular type**—it is associated with embedded tooth
 - **Extrafollicular type**—it is not associated with embedded tooth.

Clinical Features

- **Incidence**—it represents 3% of odontogenic tumors and is a developmental outgrowth of odontogenic tissue.



Fig. 51-4: Swelling seen in anterior maxillary region due to adenomatoid odontogenic tumor or cyst.

- *Age and sex distribution*—it is found in individuals ranging from 5 to 50 years with 70% occurrence in 2nd decade with an average age of 16 years. Females are affected more than males in a ratio of 2:1.
- *Site*—it occurs more commonly in the maxilla than in the mandible, usually in the anterior region and especially in the cuspid area (**Fig. 51-4**). It is commonly associated with an unerupted tooth.
- *Symptoms*—commonly presented as an area of swelling over an unerupted tooth which is asymptomatic. Sometimes, it may expand cortical bone but is not invasive.
- *Signs*—it is frequently associated with an unerupted tooth in which the epithelial proliferation is confined within a connective tissue capsule that is attached to the tooth in a manner similar to the attachment of a dentigerous cyst. When the tumor occurs independently of unerupted teeth, it is often encapsulated. The tumor causes expansion of bone and fluctuation may be elicited.
- *Progress*—it is slow growing and is a gradually increasing painless swelling leading to asymmetry, frequently associated with missing teeth.
- *Extraosseous tumor*—very uncommonly, it develops extraosseously usually in the gingiva.

Radiographic Features

- *Radiodensity*—well-demarcated mixed radiolucent or opaque lesion.
- *Site*—tumor surrounds the entire tooth, most often canine in the maxilla. Radiolucency usually extends apically beyond the cemento-enamel junction.
- *Margin*—it may or may not be well circumscribed. Borders are sclerotic.
- *Internal structure*—unilocular radiolucency but may contain faint to dense radiopaque foci which may be seen peripherally as the lesion matures (**Fig. 51-5**). Dense cluster of radiopacities appear as 'small pebbles'.

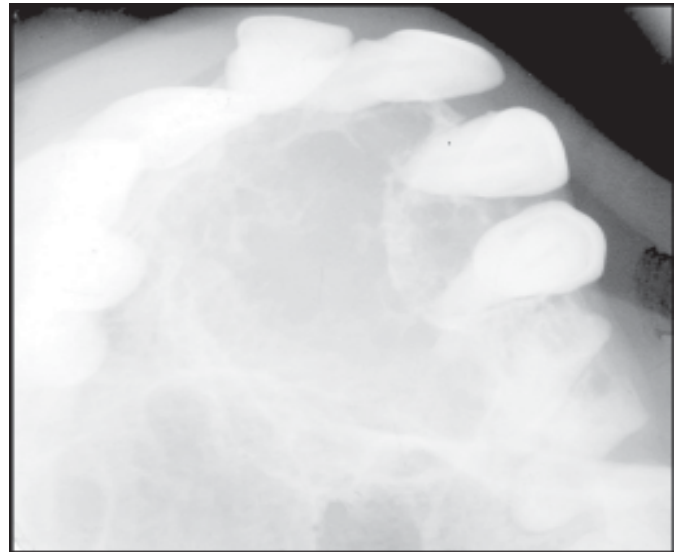


Fig. 51-5: Radiolucency showing calcification in it may be due to AOT or AOC.

- *Target appearance*—there is a radiolucent circumferential halo which envelops a dense, central and round radiodense mass.
- *Effect on surrounding structure*—separation of roots or displacement of an adjacent tooth occurs frequently. There is also cortical expansion and root resorption. The lesion may inhibit the eruption of involved tooth (**Fig. 51-6**).

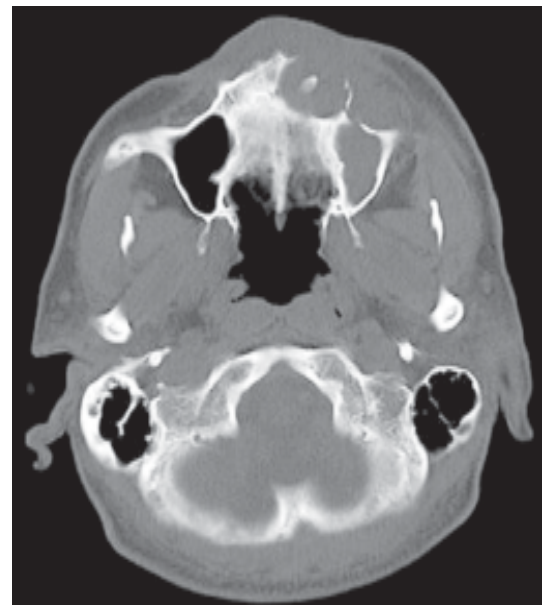


Fig. 51-6: CT scan of AOT or AOC showing lesion with impacted tooth in maxilla (Courtesy Dr Iswar).

Differential Diagnosis

- *Dentigerous cyst*—it is seen in 2nd to 4th decade as compared to adenomatoid odontogenic tumor which is seen in young age. It is seen in posterior region as

compared to adenomatoid odontogenic tumor which is seen in anterior region. Adenomatoid odontogenic tumor has tendency to surround more than just crown of the unerupted tooth.

- *Ameloblastoma*—it is more common in older age and posterior region. It is multilocular.
- *Ameloblastic fibroma*—it is seen in premolar-molar region and is multilocular.
- *Ameloblastic fibro-odontoma*—it is multilocular and radiopacities of enamel and dentin are seen inside the radiolucency as compared to adenomatoid odontogenic tumor where snowflakes are seen at the periphery.
- *Calcifying odontogenic cyst*—it occurs in older age as compared to adenomatoid odontogenic tumor and mandibular premolar area is mostly affected.
- *Odontogenic fibroma or myxoma*—tennis racket pattern is seen.

Why Discarded and Updated Term

Today we call this tumor as an adenomatoid odontogenic cyst. The reason for this is that after close inspection of the cyst's it seems proliferation which fills the lumen emerges from the epithelial lining. The calcification present in this cyst is identified as dentinoid material which derived from root sheath epithelium to induce root dentin. So due to all these factors more appropriate term for adenomatoid odontogenic tumor will be adenomatoid odontogenic cyst.

Cementoma

It is a true neoplasm of functional cementoblasts, which forms large masses of cementum or cementum-like tissue on the tooth root.

Clinical Features

- *Age and sex*—it occurs most frequently under the age of 25 years and with no significant sex predilection.
- *Site*—mandible is affected three times more frequently than the maxilla. Mandibular first molar is the most frequently affected tooth (**Fig. 51-7**); other involved teeth are the mandibular second and third molars.
- *Symptoms*—associated tooth is vital unless coincidentally involved. In some cases, pain may be there.
- *Sign*—lesion is slow growing and may cause expansion of cortical plates of bone. Periapical cementomas are multiple.

Radiographic Features

- *Appearance*—there is an area of increased density surrounded by the dark line of the fibrous capsule and with a thin white line of the adjacent cortical layer of the bone.



Fig. 51-7: Cementoma presenting as radiopaque structure in relation with lower left first molar.

Why Discarded and Updated Term

Nowadays this disease is recognized as periapical cemento-osseous dysplasia. This is disorganized product of bone periodontal membrane cementum complex.

True Cementoma

Earlier students were told the true cementoma represents benign proliferation of cementoblasts present in roots of premolar and molar teeth.

Why Discarded and Updated Term

Nowadays, this disease is recognized as cementoblastoma which is slow growing neoplastic proliferation of cementoblasts.

Gigantiform Cementoma

This is rare case of large round radiopaque mass seen in jaws.

Why Discarded and Updated Term

Nowadays this disease is recognized as ossifying fibroma. The reason for this is that these lesions are usually large ossifying fibroma with mature ossification.

Reparative Giant Cell Granuloma

It occurs in young persons usually before the age of 20 years. It is most often seen anterior to the first permanent molar. Most of the lesions are single, but more than one lesion can be seen. Spontaneous regression can be seen in reparative granuloma.

Size of the lesion varies greatly from a very small localized area of bone destruction, to one involving greater part of the bone. The margins may be poorly defined, so that exact limits of the lesion are unclear and indefinite. There is no cortex at the periphery of the lesion, as seen with the cyst. The outer and inner cortical plate of bone may become involved in the extension of the tumor and become destroyed, but the periosteum usually lays down a bony covering. The jaw is said to be expanded by the tumor. There is undulating or irregular bony covering.

Why Discarded and Updated Term

Nowadays, this is called central giant cell tumor. The reason for this is that the process which occurs in this disease is destructive rather than reparative. Second reason is that it is not a granuloma but proliferative reaction.

Aneurysmal Bone Cyst

It is an uncommon hemorrhagic lesion of the bone which is rarely seen in the jaw. It was first described as a clinicopathological entity by Jaffe and Lichtenstein in 1942. It is most often categorized as to be a tumor-like reactive lesion of bone. The name of this entity is misleading, in that, it does not contain vascular aneurysms and it is not a true bony cyst.

It represents an exaggerated localized proliferative response of the vascular tissue. It may be similar to peripheral and central giant cell granuloma to which it resembles in some ways including the histological presence of giant cells.

Pathogenesis

- *Local alteration in hemodynamics*—persistent local alteration in hemodynamics leads to increased venous pressure and development of dilated and engorged vessels in transformed bone area. Resorption of bone occurs, to which giant cells are related and this is replaced by connective tissue, osteoid and new bone.
- *Repair of hematoma of bone*—exuberant attempt at repair of hematoma of bone, similar to that of central giant cell granuloma. But, in the case of aneurysmal bone cyst, it is postulated that hematoma maintains a circulating connection with the damaged vessel. This would lead

to a slower flow of blood through the lesion and account for a clinical “welling” of blood. This is the only difference between the aneurysmal cyst and the giant cell granuloma that is in later lesion, the blood vessels fail to retain circulating connection with lesion.

- *Hemodynamic force*—Biesecker and his associates have proposed a new hypothesis for etiology and pathogenesis of this lesion that a primary lesion of the bone initiates an arteriovenous fistula and thereby creates, via its hemodynamic forces, a secondary reactive lesion of the bone. Thus occurrence of this cyst is secondary in association with osseous lesions like non-osteogenic fibroma, benign osteoblastoma and hemangioma of bone.

Clinical Features

- *Age*—although it may be seen in adults, it is more commonly seen as abnormalities of older children and adolescents with more than 90% of lesions occurring in individuals younger than the age of 30 years.
- *Sex*—it is more common in females than in males.
- *Site*—it usually involves the mandibular molar region as compared to anterior region.
- *History*—history of traumatic injury and of recent displacement of teeth which remain vital.
- *Symptoms*—aneurysmal cyst of the jaw produces a firm swelling which may be painful and tender on motion. Swelling becomes progressively worse and the rate of development is often described as rapid. Sometimes, patient may complain of difficulty in opening the mouth when there is impingement of the lesion on the capsule of TMJ.
- *Signs*—usually there is tilting or bodily displacement of teeth in the affected areas though it does not devitalize the affected tooth. Excessive bleeding may occur. When the lesion perforates the cortex and is covered by periosteum or only a thin shell of bone, it may exhibit springiness or egg-shell crackling, but it is not pulsatile. Bruit is not heard.

Radiographic Features

- *Radiodensity*—the aneurysmal bone cyst is an expansile osteolytic process within the affected bone and is projected as a definite radiolucency.
- *Internal structure*—invariably, fine septa are seen crossing through the lesion in a random pattern.
- *Appearance*—the term ‘soap bubble’ may be applied to describe an occasional multilocular radiographic appearance.
- *Margins*—are somewhat less regular and distinct than odontogenic cyst but more discrete than a central malignancy.

- *Expansion*—as the cyst enlarges to more than a few centimeters in anterior-posterior dimension, it also produces expansion of the buccal and lingual cortical plates.
- *Teeth*—simple tilting and bodily displacement of erupted teeth. Some degree of external root resorption may be seen though it will not devitalize the tooth.
- *Cortex*—there is also buccal and lingual expansion of the cortex often marked and described as ballooning or blowing out.

Differential Diagnosis

- *Ameloblastoma*—it is more common in older age group and in the mandibular posterior region.
- *Giant cell granuloma*—more common in the anterior region.
- *Central hemangioma*—it is more common in the mandible and it shows profuse hemorrhage if aspirated. Bruit is heard in such lesion.
- *Multilocular cyst*—it is more common in the mandibular posterior region of jaw and also the borders of the lesion are well defined.
- *Odontogenic myxoma*—it is more frequently associated with congenitally missing or unerupted tooth. It shows a typical honeycomb appearance.
- *Cherubism*—it occurs in a younger age group and it is a bilateral lesion.
- *Metastatic tumor*—it occurs in older age group.
- *Giant cell lesion of hyperparathyroidism*—more common in the older age group. Serum shows high levels of alkaline phosphatase.

Why Discarded and Updated Term

Today we call this cyst as central giant cell tumor. The reason for this is that all the central giant tumors have venous pressure bleeding quality. ABC has macroscopic blood-filled spaces rather than microscopic spaces. The term aneurysmal is also false as blood-filled spaces contain fibroblasts rather than endothelium. It also lacks cystic lining so the term aneurysmal bone cyst is discarded.

Eagle Syndrome

In this syndrome there is neck pain associated with elongated calcified styloid process.

Why Discarded and Updated Term

Today we called this as part of DISH syndrome (diffuse intraosseous skeletal hypertrophy). Initially, it is thought

to be occurred due to calcification of stylohyoid ligament. But nowadays it is seen that there is actually ossification of stylohyoid ligament with mature bone. Also, there is evidence that maximum of intervertebral ligament are also ossified.

Hand-Schuller-Christain Disease, Letterer Siwe Disease, Eosinophilic Granuloma and Histiocytosis X

It is an inflammatory reticuloendothelioma condition with evidence suggesting that it may be reaction to some type of infection. There is pathological accumulation of histiocytes and eosinophilic leukocytes.

Why Discarded and Updated Term

Nowadays dominant histolytic cells identified as antigen processing Langerhans cells. So it is now called Langerhans cell histiocytosis (LCH). All the three diseases are now called separated clinical entity under the heading of Langerhans cell histiocytosis.

Neurilemmoma, Neurinoma and Neuro-lemmoma

The above terms are derived from Greek word: neuron meaning nerve and lemma means husk or covering.

Why Discarded and Updated Term

Today we called this as Schwannoma. The reason for this is that tumor consists of Schwann cells around the nerve and not the nerve fiber or nerve sheath. Also, there is only proliferation of Schwann cells and not the basal lamina or other component of the neurilemma.

Traumatic Bone Cyst

It is empty space seen in the bone and many times it is not related to trauma.

Why Discarded and Updated Term

Today we call this as idiopathic bone cavity. There is no epithelium present in this cavity so use of term traumatic bone cyst should be avoided.

Suggested Reading

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5. Marx RE, Stern DS. Oral and maxillofacial pathology, a rationale for Diagnosis and treatment. Quintessence Publishing Co, Inc 2003.
6. Krandorf MJ, Sweet DE. Aneurysmal bone cyst: concept, controversy, clinical presentation and imaging. AM J Roentgel 195;164:573-80.

Appendices

1

Causes and Classifications

Classifications of Diseases

White Lesions

Variation in structure and appearance of normal mucosa

- Leukoedema
- Fordyce's granules
- Linea alba

White lesion with definite precancerous potential

- Leukoplakia
- Erythroplakia
- Tobacco keratosis, actinic keratosis
- Lesion associated with electrogalvanism
- Carcinoma *in situ*
- Verrucous carcinoma
- Lichen planus
- Lichenoid reaction
- Oral submucous fibrosis
- Dyskeratosis congenita
- Lupus erythematosus
- Acanthosis nigricans

White lesion without precancerous potential

- Traumatic keratosis
- Focal epithelial hyperplasia
- Psoriasis
- Geographic tongue
- Pachyonychia congenita
- White sponge nevus
- Hereditary benign epithelial dysplasia
- Stomatitis nicotina
- Hyperkeratosis palmoplantaris with gingival hyperkeratosis
- Darrier's disease
- Intraoral skin grafts
- Pseudoxanthoma elasticum
- Hyalinosi cutis et mucosae oris
- Oral condyloma acuminatum
- Hairy leukoplakia

Nonkeratotic white lesion

- White hairy tongue

- Burns –thermal, chemical, electrical
- Desquamative gingivitis
- Epidermolysis bullosa
- Pemphigus
- Benign mucous membrane pemphigoid (BMMP)
- Diphtheria
- Syphilitic mucus patches
- Acute necrotizing ulcerative gingivitis (ANUG)
- Koplik's spot of measles
- Candidiasis
- Median rhomboidal glossitis
- Verruca vulgaris

Precancerous Lesion and Condition

Precancerous lesion

- Leukoplakia
- Erythroplakia
- Palatal lesion associated with reverse smoking
- Verrucous hyperplasia
- Carcinoma *in situ*

Precancerous condition

- Oral submucous fibrosis
- Sideropenic anemia
- Erosive lichen planus
- Discoid lupus erythematosus
- Dyskeratosis congenita

Histological Classification of Cancer and Precancer of the Oral Mucosa—Histological typing of cancer and precancer (WHO 1997)

Carcinomas

- Squamous cell carcinoma
- Verrucous carcinoma
- Basaloid squamous cell carcinoma
- Adenoid squamous cell carcinoma
- Spindle cell carcinoma
- Adenosquamous carcinoma
- Undifferentiated carcinoma

Benign lesions capable of microscopically resembling oral squamous cell carcinoma and oral verrucous carcinoma

- Papillary hyperplasia
- Granular cell tumor
- Discoid lupus erythematosus
- Median rhomboid glossitis
- Keratoacanthoma
- Necrotizing sialometaplasia
- Juxtaoral organ of Chievitz
- Chronic hyperplastic candidiasis
- Verruciform xanthoma
- Verruca vulgaris
- Condyloma acuminatum

Precancerous lesions (Clinical Classification)

- Leukoplakia
- Erythroplakia
- Palatal keratosis associated with reverse smoking

Precancerous lesions (Histological Classification)

- Squamous epithelial dysplasia
- Squamous cell carcinoma *in situ*
- Solar keratosis

Benign lesions capable of resembling oral precancerous lesions

- White lesions resembling leukoplakia
- Red lesions resembling erythroplakia
- Focal epithelial hyperplasia
- Reactive and regenerative atypia

Precancerous conditions

- Sideropenic dysphagia
- Lichen planus
- Oral submucous fibrosis
- Syphilis
- Discoid lupus erythematosus
- Xeroderma pigmentosum
- Epidermolysis bullosa

Red Lesion of Oral Mucosa**Inflammatory condition****Inflammation associated with traumatic injury**

- *Mechanical*—cheek biting, ill fitting denture, sharp edge, overhanging restoration and ecchymoses due to trauma.
- *Chemical*—aspirin, formocresol and TCA.
- *Thermal*—hot food, hot beverage and hot instrumentation.
- *Radiation*—mucositis, cheilitis.

Infection**Bacterial**

- Scarlet fever (red fever)
- Gonococcal stomatitis
- Vincent infection or gingivostomatitis
- Acute pharyngitis, tonsillitis

Fungal

- Atrophic candidiasis
- Angular cheilitis
- Acute pseudomembranous candidiasis

Viral

- Measles—Koplik's spot
- Lymphnodular pharyngitis
- Herpes simplex infection
- Herpes zoster
- Herpangina
- Chickenpox
- Hand-foot-mouth disease

Allergic/Immunological

- Pyogenic granuloma
- Giant cell epulis
- Pregnancy tumor
- Traumatic hemangioma
- Inflammatory fibrous hyperplasia
- Desquamative gingivitis

Congenital-hereditary developmental condition

- Hemangioma
- Sturge-Weber syndrome
- Rendu-Osler-Weber disease
- Hereditary hemorrhage telangiectasia
- Median rhomboidal glossitis
- Geographic tongue
- Melkersson-Rosenthal syndrome
- Crohn's disease

Vascular disease and blood disorder

- Purpura
- Polycythemia
- Agranulocytosis
- Polyarteritis nodosa
- Leukemia

Dermatological

- Pemphigus
- Erythema multiforme
- Stevens Johnson
- Lichen planus
- Lichenoid reaction
- Epidermolysis bullosa
- Psoriasis
- Lupus erythematosus

Other systemic disease

- Uremic stomatitis
- Diabetes stomatitis
- Scurvy
- Pernicious anemia
- Ulcerative stomatitis

Premalignant and malignant lesion

- Atrophic leukoplakia
- Erythroplakia
- Squamous cell carcinoma
- Carcinoma *in situ*
- Kaposi's sarcoma
- Hemangioendothelioma

Vesiculobullous Lesions**1st Classification***Hereditary*

- Epidermolysis bullosa
- Familial benign chronic pemphigus
- Dyskeratosis congenita
- Acrodermatitis enteropathica

Viral infection

- Primary herpetic gingivostomatitis
- Secondary herpetic gingivostomatitis
- Varicella (chickenpox)
- Shingles (herpes zoster)
- Measles
- Hand-foot-mouth disease
- Small pox and cat-scratch disease
- Infectious mononucleosis
- Herpangina
- Acute lymphnodular pharyngitis
- AIDS

Mucocutaneous diseases

- Pemphigus vulgaris
- Pemphigus vegetans
- Benign mucous membrane pemphigoid
- Bullous pemphigoid
- Lichen planus
- Toxic epidermal necrolysis

Miscellaneous

- Oral submucous fibrosis
- Hyperacidity or gastritis
- Constipation and malabsorption syndrome
- Impetigo
- Oral blood blister
- Erythema multiforme

2nd Classifications*Vesicular lesion**Non-febrile lesion*

- Recurrent herpes labialis
- Recurrent herpes stomatitis
- Reiter's syndrome
- Contact stomatitis
- Impetigo
- Dyskeratosis congenita

Febrile lesion

- Herpetic gingivostomatitis
- Herpangina
- Hand-foot-mouth disease
- Chickenpox
- Herpes zoster
- Smallpox

Bullous lesion

- Pemphigus
- Familial benign chronic pemphigus
- Bullous pemphigoid
- Benign mucous membrane pemphigoid
- Epidermolysis bullosa
- Dermatitis herpetiformis

- Erythema multiforme
- Stevens-Johnson syndrome

Cysts of the Oral Cavity*Odontogenic**Developmental*

- Primordial cyst
- Odontogenic keratocyst
- Gingival cyst of infant
- Gingival cyst of adult
- Lateral periodontal cyst
- Dentigerous cyst
- Eruption cyst
- Calcifying epithelial odontogenic cyst

Inflammatory

- Radicular cyst
- Periodontal cyst
- Inflammatory collateral cyst
- Paradental cyst

Non-odontogenic

- Nasopalatine cyst
- Median palatine cyst
- Median alveolar cyst
- Median mandibular cyst
- Globulomaxillary cyst
- Nasolabial cyst
- Nasoalveolar cyst
- Simple bone cyst
- Hemorrhagic bone cyst

Cysts Associated with maxillary sinus

- Benign mucosal cyst of maxillary antrum
- Mucocele–mucus retention cyst
- Surgical ciliated cyst

Soft tissue cysts

- Dermoid
- Epidermoid
- Branchial
- Thyroglossal
- Anterior median lingual cyst
- Oral cyst with gastric epithelium
- Cystic hygroma
- Parasitic cyst, hydatid cyst, cysticercosis cellulosae

Cysts of the salivary gland

- Mucocele
- Ranula

Oral Manifestation of AIDS*Fungal infection*

- Candidiasis
- Histoplasmosis
- Toxoplasmosis
- Cryptococcosis
- Geotrichosis

- Mucormycosis
- Aspergillosis
- Actinomycosis

Bacterial

- HIV gingivitis
- HIV necrotizing gingivitis
- HIV periodontitis
- Sinusitis
- STD

Viral

- Herpes simplex
- Herpes zoster
- Hairy leukoplakia
- Verruca vulgaris
- Condyloma acuminatum
- Focal epithelial hyperplasia
- Cytomegalovirus infection

Neoplasm

- Kaposi's sarcoma
- Squamous cell carcinoma
- Non-Hodgkin's lymphoma

Neurological

- Trigeminal neuralgia
- Facial palsy
- AIDS dementia

Miscellaneous

- Recurrent aphthous stomatitis
- Erythema multiforme
- Lichenoid reaction
- Progressive necrotizing ulceration
- Toxic epidermal necrolysis

Odontogenic Tumor

1st Classification

Benign odontogenic tumor

Odontogenic epithelium without odontogenic ectomesenchyme

- Ameloblastoma
- Squamous odontogenic tumor
- Pindborg tumor
- Clear cell odontogenic tumor

Odontogenic epithelium with odontogenic ectomesenchyme with or without dental hard tissue formation

- Ameloblastic fibroma
- Ameloblastic fibro-odontoma
- Ameloblastic fibrodentinoma
- Odontoameloblastoma
- Adenomatoid odontogenic tumor
- Complex and compound odontome

Odontogenic ectomesenchyme with or without included odontogenic epithelium

- Odontogenic fibroma
- Odontogenic myxoma
- Benign cementoblastoma

Malignant odontogenic tumor

Odontogenic carcinoma

- Malignant ameloblastoma
- Primary intraosseous carcinoma
- Malignant variant of other odontogenic epithelial tumor
- Malignant changes in odontogenic cyst

Odontogenic sarcoma

- Ameloblastic fibrosarcoma
- Ameloblastic dentinosarcoma

2nd Classification

Benign

Epithelial

- *With inductive changes in the connective tissue*
 - AOT
 - Ameloblastic fibroma
 - Dentinoma
 - Calcifying odontogenic tumor
 - Odontoameloblastoma
 - Odontoma
- *Without inductive changes in connective tissue*
 - Ameloblastoma
 - CEOT
 - Epithelial atypia
 - Ameloblastic changes in odontogenic cyst

Mesenchymal

- Odontogenic myxoma
- Odontogenic fibroma
- Cementoma
- Periapical cemental dysplasia
- Cementifying fibroma
- Benign cementoblastoma

Malignant

Epithelial

- *With inductive changes in connective tissue*
 - Ameloblastic fibrosarcoma
 - Ameloblastic odontosarcoma
- *Without inductive changes in connective tissue*
 - Malignant ameloblastoma
 - Primary intraosseous carcinoma
 - Malignant changes in odontogenic cyst

Benign Tumors of the Jaw

Odontogenic tumors

Epithelial

- Ameloblastoma
- Adenoameloblastoma
- Enameloma
- Pindborg tumor (CEOT)

Mesenchymal

- Dentinoma
- Cementoma
- Cementoblastoma

Mixed

- Ameloblastic fibroma
- Ameloblastic fibro-odontoma
- Ameloblastic odontoma

- Odontogenic myxoma
- Compound composite odontoma
- Complex composite odontoma
- Odontogenic fibroma

Developmental

- Dense invaginatus or dilated odontome
- Dense evaginatus

Non-odontogenic tumors

Epithelial tissue

- Papilloma
- Keratoacanthoma
- Adenoma

Fibrous connective tissue

- Fibroma
- Myxoma
- Fibrous hyperplasia
- Fibrous epulis
- Fibrous histiocytoma

Cartilage

- Chondroma
- Chondroblastoma
- Chondromyxoid fibroma

Adipose tissue

- Lipoma

Bone

- Osteoma
- Osteoid osteoma
- Osteoblastoma
- Exostosis and tori- torus palatinus, torus mandibularis
- Enostosis

Vascular tissue

- Hemangioma
- Lymphangioma
- Glomus tumor
- Hemangiopericytoma

Neural tissue

- Neurofibroma
- Neurilemmoma
- Schwannoma
- Ganglioneuroma
- Traumatic neuroma
- Melanotic neuroectodermal tumor of infancy

Muscle

- Leiomyoma
- Rhabdomyoma
- Myoblastoma

Giant cell tumors

- Central and peripheral giant cell tumor
- Giant cell granuloma
- Giant cell reparative granuloma
- Giant cell tumor of hyperthyroidism

Teratoma

Salivary gland tumors

- Adenoma

- Oncocytoma
- Oxyphilic adenoma
- Warthin's tumor
- Pleomorphic adenoma

Classification of Tumor

	<i>Tissue of origin</i>	<i>Benign</i>	<i>Malignant</i>
Epithelial tumors	Squamous epithelium	Squamous cell papilloma	Squamous cell carcinoma
	Transitional epithelial	Transitional cell papilloma	Transitional cell carcinoma
	Glandular epithelium	Adenoma	Adenocarcinoma
	Basal cell layer	—	Basal cell carcinoma
	Neuroectodermal	Nevus	Melanoma
	Hepatocyte	Liver cell adenoma	Hepatocellular carcinoma
Mesenchymal	Adipose tissue	Lipoma	Liposarcoma
	Adult fibrous tissue	Fibroma	Fibrosarcoma
	Embryonic fibrous tissue	Myxoma	Myxosarcoma
	Cartilage	Chondroma	Chondrosarcoma
	Bone	Osteoma	Osteosarcoma
	Synovium	Benign synovioma	Synovial sarcoma
	Skeletal muscle	Rhabdomyoma	Rhabdomyosarcoma
	Smooth muscle	Leiomyoma	Leiomyosarcoma
	Mesothelium		Mesothelioma
	Blood vessels	Hemangioma	Angiosarcoma
	Lymph vessels	Lymphangioma	Lymphangiosarcoma
	Glomus cell	Glomus tumor	
	Meninges	Meningioma	Invasive meningioma
	Hemopoietic cell		Leukemia
	Lymphoid tissue		Malignant lymphoma
	Nerve sheath	Neurilemmoma	Neurogenic sarcoma
	Nerve cell	Ganglioneuroma	Neuroblastoma
Mixed tumors	Salivary gland	Pleomorphic adenoma	Malignant salivary gland tumor
Tumor of more than one germ cell layer	Totipotent cells in gonads or in embryonal rests	Mature teratoma	Immature teratoma

Malignant Tumor of the Jaw

Odontogenic tumor

- Ameloblastic sarcoma and Fibrosarcoma
- Odontogenic sarcoma
- Carcinoma in odontogenic cyst

Non-odontogenic tumor

Epithelial

- Squamous cell carcinoma
- Mucoepidermoid carcinoma
- Adenocarcinoma
- Basal cell carcinoma
- Transitional cell carcinoma
- Melanoma
- Verrucous carcinoma
- Intraepidermoid carcinoma

Fibrous connective tissue

- Fibrosarcoma

Adipose tissue

- Liposarcoma

Cartilage

- Chondrosarcoma

Bone

- Osteosarcoma
- Osteochondrosarcoma

Vascular

- Hemangioendothelioma

Neural tissue

- Neurosarcoma
- Neurofibrosarcoma

Muscle

- Leiomyosarcoma
- Rhabdomyosarcoma

Lymphoid tissue

- Hodgkin and non-Hodgkin lymphoma
- Lymphosarcoma
- Reticular cell sarcoma
- Ewing sarcoma
- Burkitt's lymphoma
- Multiple myeloma
- Leukemia

Tumor of salivary gland

- Mucoepidermoid carcinoma
- Adenocystic carcinoma
- Adenocarcinoma
- Acinic cell carcinoma
- Malignant change in pleomorphic adenoma

Granulomatous Diseases**Specific or infective type**

- Syphilis
- Yaws and Bejel
- Tuberculosis
- Leprosy
- Melkersson Rosenthal syndrome
- Heerfordt's syndrome
- Blastomycosis
- Coccidioidomycosis
- Cryptococcosis
- Rhinosporidiosis
- Pyostomatitis vegetans
- Actinomycosis
- Candidiasis
- Aspergillosis
- Mucormycosis
- Lymphogranuloma venereum
- Granuloma inguinale

Non-specific

- Periapical granuloma
- Pyogenic granuloma
- Peripheral and central giant cell granuloma
- Foreign body granuloma
- Traumatic granuloma
- Crohn's disease
- Pulsating granuloma

Histiocytosis**Non-lipid**

- Eosinophilic granuloma
- Hand-Schüller-Christian disease
- Letterer-Siwe disease

Lipid

- Gaucher's disease
- Niemann-Pick disease

Malignant

- Wegener's granulomatosis
- Midline lethal granuloma
- Hodgkin's disease

Gingival Hyperplasia (Clinical Classification)**Localized**

- Marginal
- Papillary
- Discrete

Generalized

- Diffuse
- Marginal
- Papillary

Gingival Hyperplasia (Etiological Classification)**Inflammatory****Acute**

- Gingival or periodontal abscess
- Acute exacerbation of chronic inflammatory enlargement

Chronic**Local irritants**

- Calculus
- Food lodgment area
- Occlusal trauma
- Mouth breathing
- Overhanging restoration
- Foreign bodies
- Poor oral hygiene

Infection

- Bacterial
- Viral
- Fungal
- Fusospirochetal
- Parasitic
- Saprophytic

Allergy**Non-inflammatory**

- Familial or hereditary
- Developmental
- Congenital
- Idiopathic
- Drug induced
- Dilantin sodium
- Phenobarbitone
- Nifedipine
- Cyclosporine

Combined**Conditional**

- Hormonal (puberty and thyroid dysfunction)
- Leukemic enlargement—myelogenous or monocytic
- Polycythemia vera
- Vitamin C deficiency
- Non-specific conditional enlargement
 - Epulis—fibrous
 - Epulis fissuratum
 - Eosinophilic granuloma
 - Plasma cell granuloma

- Crohn's disease
- Peripheral giant cell granuloma
- Wegener's granulomatosis

Cyst

- Eruption
- Gingival

Neoplastic

Benign

- Fibroma
- Granular cell myoblastoma
- Papilloma
- Hemangioma
- Peripheral ameloblastoma
- Peripheral odontogenic fibroma

Malignant

- Squamous cell carcinoma
- Fibrosarcoma
- Reticular cell sarcoma
- Melanoma
- Malignant lymphoma
- Malignant hemangioendothelioma or hemangioendotheliosarcoma
- Metastasis from lung, colon, testis, kidney and cervix

Syndrome associated with gingival hyperplasia

- Rutherford
- Cannon's disease
- Cross
- Zimmermann-Laband
- Cowden's
- Papillon Lefevre
- Tuberous sclerosis
- Sturge-Weber
- Melkersson-Rosenthal

Oral Pigmentation

1st Classification

Exogenous

Occupational

- Lead industry—lead
- Match industry—phosphorus
- Fluorescent lamp industry—mercury
- Photography—silver

Habits

- Tobacco—in the form of smoking, chewing and snuff dipping
- Pan—catechu, pan and pan masala
- Food—food stuff, black berries, blue grapes, coloring agent of sweets and chocolates

Therapeutic

- Drugs—anti-malarial drugs and contraceptives
- Metallic salts—gold for tuberculosis, bismuth for diarrhea, silver for nasal drop, mercury for diuretic

Others

- Amalgam tattoos
- Other self made tattoos—graphite, methylene blue

- Black hairy tongue
- Poor oral hygiene

Hypopigmentation and de-pigmentation

- Albinism
- Vitiligo
- Pernicious anemia
- Hormonal disturbances
- Parasitic infection
- Chédiak-Higashi syndrome
- Tuberous sclerosis
- Cross syndrome
- Severe burns
- Extensive traumatic injury

Endogenous

Physiologic

- Ethnic, racial variation
- Physiologic melanotic macule and papule
- Fordyce's granules
- Postmenopausal changes in sex hormones
- Pregnancy

Pathological

- Addison's disease
- Acromegaly
- Peutz-Jeghers syndrome
- Lentigo
- Nevi (intradermal, compound, junctional, blue)
- Neurofibromatosis
- Metastatic carcinoma
- Malnutrition
- *Acanthosis nigricans*
- Cyanosis
- Hemolytic anemia
- Jaundice
- Hemochromatosis
- Hemosiderosis
- Porphyria
- Carotenemia
- Hematoma-ecchymoses-varicosities-purpura
- Niemann-Pick disease
- Diabetic melanosis
- Simmond's disease

2nd Classification

Benign melanocytic lesions

- Racial pigmentation
- Physiologic pigmentation
- Smoking associated pigmentation
- Ephelis
- Lentigo
- Oral melanotic macules
- Nevi
- Hematoma

Neoplastic conditions

- Melanoma
- Multiple neurofibromatosis
- Neuroectodermal tumor of infancy
- Hemangioma
- Kaposi's sarcoma

Different type of pigmentation and their features				
Condition	Appearance	History	Age	Frequency
Melanoplakia	Macule of varying size, shape and location of oral mucosa	No symptoms	Birth to 1 year	Common in dark race
Amalgam tattoo	Macule on gingiva or edentulous ridge	Amalgam filled teeth	5 years and older	Common
Petechiae and ecchymosis	Macule anywhere in oral cavity or skin	Recent trauma bleeding diathesis	Young children	Occasional
Melanotic macule	Small macule < 2 cm on lower lip gingiva or buccal mucosa	—	41-42 years	M/F 1:2.2
Superficial spreading melanoma	Enlarging macule on palate or maxillary sinus	Several years Slowly enlarging	Mean age 50.5	Rare M/F=2:1
Junctional nevus	Macule occurs anywhere in oral cavity		Mean age 38	Rare in oral cavity
Heavy metal poisoning	Macular lesion— free gingiva	Malaise anemia	Working age	Rare
Peutz-Jeghers syndrome	Macule around lip buccal mucosa, finger body orifice	Melanin intestinal polyposis	Distinct at puberty	Rare
Albright syndrome	Café au lait spot on skin and mucous membrane	Skeletal endocrine	—	—
Firm lesions Amalgam fragment	Macule on gingiva or edentulous ridge	Firm mass in tissue	5 years and older	Common
Late hematoma	Mass and swelling	Previous trauma	—	Occasional
Giant cell granuloma	Moderately firm macule and nodule on gingiva and alveolar	Slowly expanding	30 years and over	Occasional
Pigmented lesion	Papule nodule or polypoid mass usually on buccal mucosa	Present for some time	—	Occasional
Intramucosal nevus	Papule nodule or polypoid mass	Present from birth	—	Rare
Compound nevus	Same	Same	—	Same
Melanoma	Pigmented or amelanotic nodule polypoid mass on palate and maxillary gingiva	Rapidly enlarging	—	Rare M/F 2:1
Neuroectodermal tumor	Expansion of labial sulcus pigmented or pink	Slowly expanding mass in anterior maxilla	Birth to 1 year	Rare

Pigmentation due to exogenous deposits

- Amalgam tattoo
- V Graphite tattoo
- Heavy metal pigmentation
- Drug induce pigmentation

Miscellaneous condition

- Peutz-Jeghers syndrome
- Addison disease
- Pigmented *lichen planus*
- Hairy tongue
- Mucocele
- HIV-associate oral pigmentation
- Endocrinopathic pigmentation

Diseases of the Maxillary Sinus**Traumatic**

- Fracture of maxilla, nasal bone, zygoma and orbital floor
- Hematoma
- *Foreign bodies*—root piece, bullet injury, antroliths
- Oroantral fistula

Inflammatory

- Acute and chronic sinusitis
- Local hyperplasia from odontogenic infection
- Antral polyp
- Osteomyelitis

Cysts**Intrinsic**

- Mucus retention cyst (mucocele)
- Serous cyst
- Pseudo cyst

Extrinsic

- *Odontogenic*
 - Radicular
 - Dentigerous
 - Primordial
 - Keratocyst
- *Non-odontogenic*
 - Globulomaxillary
 - Traumatic
 - Aneurysmal bone cyst (ABC)

Neoplasm**Odontogenic**

- Ameloblastoma
- Adenoameloblastoma
- Calcifying Epithelial Odontogenic Tumor (CEOT)

Nonodontogenic

- Exostosis
- Enostosis

Malignant

- Squamous cell carcinoma
- Midline lethal granuloma*

Metabolic

- Fibrous dysplasia
- *Leontiasis ossea*

* It is controversial issue to consider midline lethal granuloma as a malignancy as many investigators in spite of taking repeated biopsies have not found neoplastic tissue in these lesions.

Sexually Transmitted Diseases

- Syphilis
- Yaws and Pinta
- Gonorrhea
- Genital herpes
- Genital warts
- Nonspecific genital infection
- Trichomoniasis
- *Pediculosis pubis*
- Scabies
- *Molluscum contagiosum*
- Chancroid
- Lymphogranuloma venereum
- Lymphogranuloma inguinale
- Hepatitis B
- Cytomegalovirus inclusion body
- Infectious mononucleosis
- AIDS

Sexually Transmitted and Blood Born Disease**Bacterial and chlamydial STD (potentially curable and rarely blood borne)**

- Gonorrhea and related clinical syndrome (urethritis, cervicitis, salpingitis, pelvic inflammatory disease, disseminated systemic infection, arthritis, proctitis, pharyngitis)
- Non- and post-gonococcal urethritis and related clinical syndrome (cystitis, epididymitis, Reiter's disease, pelvic inflammatory disease)
- Syphilis and other treponemal infection (yaws pinta bejel)
- Chancroid
- Granuloma inguinale
- Lymphogranuloma venereum
- Gay bowel syndrome associated pathogen

Viral STD (currently incurable some frequently blood born)

- Herpes simplex virus infection by HSV I and II
- Infectious mononucleosis
- Cytomegalovirus infection
- Viral hepatitis
 - Hepatitis A
 - Hepatitis B
 - Hepatitis D
 - Hepatitis C
 - Hepatitis E
 - Hepatitis F
- AIDS
- T cell lymphoma and leukemia
- *Molluscum contagiosum*

Miscellaneous bacterial fungal and parasitic infection which sometime manifested as STD (usually curable and rarely blood born except candida)

- Vulvovaginal candidiasis
- Trichomoniasis
- Vaginitis, cervicitis and cystitis
- Bacterial vaginosis
- Intestinal protozoa infection
- Ectoparasitic skin infection

Other blood born infection with no evidence of sexual transmission

- Toxoplasmosis
- Malaria
- Babesiosis
- Trypanosomiasis

Autoimmune Disorder**Associated with Mucocutaneous Lesion**

- Recurrent aphthous ulcer
- Behcet's disease
- Pemphigus
- Bullous pemphigoid
- Cicatricial pemphigoid
- Dermatitis herpetiformis

Salivary gland

- Mikulicz's disease
- Sjögren's syndrome

Blood disorder

- Pernicious anemia
- Purpura

Collagen disorder

- Systemic lupus erythematosus
- Scleroderma
- Rheumatic arthritis

Miscellaneous

- Myasthenia gravis
- Dermatomyositis
- Oral submucous fibrosis

Blood Disorders**Disorders of RBC****Deficiency**

- Iron deficiency anemia
- Pernicious anemia
- Normocytic anemia
- Aplastic anemia
- Hemolytic anemia

Extra-corporeal causes

- Infection and toxin
- Hypersplenism
- Rh factor incompatibility (hemolytic disease of newborn, erythroblastosis fetalis)
- Chronic liver disease
- Autoimmune diseases
- Transfusion reaction

Intra-corporeal causes

- Hereditary spherocytosis
- Sick cell anemia
- Thalassemia
- Cooley's anemia
- Glucose-6 phosphate dehydrogenase deficiency
- Pyruvate kinase deficiency
- Folic acid and B₁₂ deficiency

Polycythemia

- Relative
- Secondary
- Vera (malignant)

Disorders of WBC**Qualitative**

- Lazy leukocyte syndrome
- Chédiak-Higashi syndrome

Quantitative

- Agranulocytosis
- Cyclic neutropenia

- Myeloblastic leukemia
- Decrease of eosinophils

Disorders of platelet**Purpura**

- Idiopathic thrombocytopenic purpura
- Secondary thrombocytopenic purpura
- Thrombotic thrombocytopenia
- Drug-associated thrombocytopenia
- Thrombocytopathia
- Glanzmann's thrombasthenia
- von Willebrand's disease
- Bernard-Soulier syndrome
- Aldrich syndrome

Thrombocytosis**Disorders of coagulation**

- Hemophilia A
- Hemophilia B
- Factor XI deficiency
- Factor X deficiency
- von Willebrand's disease

Fibro-osseous Lesions**Lesions arising from periodontal ligament**

- Periapical cemental dysplasia
- Localized fibro-osseous cemental lesion
- Florid osseous dysplasia
- Ossifying fibroma
- Cementifying fibroma

Fibrous dysplasia

- Monostotic
- Polyostotic
- Familial

Fibro-osseous neoplasm of uncertain origin or debatable origin

- Cementoblastoma
- Osteoblastoma
- Osteoid osteoma
- Juvenile ossifying fibroma
- Fibro-osteoma or osteofibroma
- Cementifying fibroma
- Paget's disease
- Ossifying fibrous epulis

Giant cell lesion

- Central giant cell tumor or central giant cell reparative granuloma
- Brown tumor of hyperthyroidism
- Aneurysmal bone cyst
- Cherubism

Mucocutaneous Disorder**Genodermatoses**

- White sponge nevus

- Darrier's disease
- Peutz-Jeghers syndrome
- Dyskeratosis congenita
- Hereditary benign intraepithelial dyskeratosis
- *Pachyonychia congenita*
- Hyalinosis cutis et mucosa oris
- *Pseudoxanthoma elasticum*

Non-infective disease

Vesicular

- Erythema multiforme
- Pemphigus
- Benign mucous membrane pemphigoid
- Bullous pemphigoid
- Epidermolysis bullosa
- Bullous Lichen planus

Non-vesicular disease

- Lichen planus
- Benign migratory glossitis

Collagen disorder

- Lupus erythematosus
- Scleroderma
- *Polyarteritis nodosa*
- Vasculitis
- Ischemic lingual necrosis
- Wegner granulomatosis
- Midline lethal granuloma

Degenerative and relative disorder

- Amyloidosis
- Oral submucous fibrosis
- Senile solar elastosis

Pigmentation

- Racial pigmentation
- Endocrinopathy
- Addison disease
- Albert syndrome
- Bronze diabetes
- Anemia

Oral Hamartomas

Odontogenic

Those involving teeth

- Dens invaginatus
- Dens evaginatus
- Talon's cusp

Those not involving teeth

- Enameloma
- Odontoma
- Gigantiform cementoma
- Dental lamina cyst of newborn

Non-odontogenic

Epithelial origin

- Epstein pearls and Bohn's nodule

- Oral and labial melanotic macule
- Pigmented cellular nevus

Vascular origin

- Hemangioma
- Lymphangioma
- Glomus tumor

Osseous origin

- Torus palatinus
- Torus mandibularis

Adipose tissue

- Lipoblastomatosis

Neural tissue

- Neurofibromatosis

Unknown or doubtful origin

- Granular cell myoblastoma
- Congenital epulis of newborn
- Melanotic neuroectodermal tumor of infancy
- Fibromatosis gingivae

Disorders of the Salivary Glands

Developmental

- Congenital aplasia or agenesis
- Congenital hypoplasia
- Atresia
- Aberrance or ectopic gland
- Diverticuli
- Accessory duct
- Congenital fistula

Inflammatory

- *Acute and chronic*
 - Staphylococcus
 - Streptococcus
 - Actinomycosis
 - Tuberculosis
- *Viral infection*
 - Mumps
 - Cytomegalovirus inclusion disease
 - Para-influenza
- Sarcoidosis
- Melkersson-Rosenthal syndrome
- Heerfordt's syndrome
- Allergy
- Secondary to sialolithiasis
- Post-irradiation to oral tumor
- Salivary fistula

Sialolithiasis

- Due to stricture of the duct
- Due to trauma or infection
- Due to salivary stone calculi
- Mucus plug

Cysts

- Mucocele
- Ranula
- Lymphoepithelial cyst
- Branchial cyst

Frey's syndrome

Sialadenosis

- Non-inflammatory swelling of salivary gland

Sjögren's syndrome**Necrotizing sialometaplasia****Neoplasms****Benign epithelial**

- Adenoma
- Oncocytoma
- Warthin's tumor
- Pleomorphic adenoma

Malignant epithelial

- Adenocarcinoma
- Mucoepidermoid carcinoma
- Adenocystic carcinoma
- Acinic cell carcinoma
- Malignant pleomorphic adenoma

Mesenchymal benign

- Hemangioma
- Lymphangioma
- Neurofibroma
- Schwannoma
- Xanthoma
- Lipoma

Malignant mesenchymal

- Rhabdomyosarcoma
- Hemangioendothelioma

Others

- Lymphoma—Hodgkin and non-Hodgkin
- Metastatic

Oral Lymphoid Tissue Lesion**Developmental lesions**

- Reactive lymphoid aggregates
- Lymphoid hamartomas
- Angio-lymphoid hyperplasia with eosinophilia

Cystic

- Lymphoepithelial cyst

Neoplastic

- Benign—Warthin's tumor
- Malignant—lymphoepithelioma (epithelial) and malignant lymphoma, Burkitt's lymphoma, non-Hodgkin lymphoma and leukemia (connective tissue)

Syndrome

- Mikulicz's disease of syndrome
- Sjögren's syndrome

Disorders of the Temporomandibular Joint**Developmental**

- Agenesis of the condyle
- Hypoplasia of the condyle
- Hyperplasia of the condyle
- Exostosis of the joint

Traumatic

- Trauma to disc
- Trauma to developing condyle or growth center
- Trauma to ligament
- Subluxation or dislocation or dislocation within displacement
- Ankylosis
- Foreign body in the joint space
- Trauma to muscle

Inflammatory**Infective**

- Pyogenic infection from staphylococci
- Syphilis
- Tuberculosis
- Actinomycosis
- Rheumatic arthritis
- Osteomyelitis
- Psoriasis
- Secondary to hepatitis B infection

Non-infective

- Still's disease
- Systemic lupus erythematosus
- Ankylosis spondylitis
- Synovitis

Non-articular arthritis

- Myositis
- Myositis ossificans
- MPDS

Metabolic Disorders

- Gout
- Chondrocalcinosis
- Oxalosis
- Amyloidosis
- Wilson's disease
- Gaucher's disease

Neoplasms**Benign**

- Osteoma
- Chondroma
- Osteochondroma
- Fibromyxoma
- Synovioma

Malignant

- Osteosarcoma
- Chondrosarcoma
- Synovial sarcoma
- Malignant lymphoma

Neuropathic

- Charcoat joint
- Ritter's sympathetic dystrophy

Internal derangement

- Disc displacement
- Disc fracture

Giant Cell Disorders**Traumatic**

- Peripheral giant cell granuloma
- Central giant cell granuloma
- Traumatic granuloma
- Epulis fissuratum
- Pyogenic granuloma
- Internal resorption
- Periapical granuloma

Infection

- Tuberculosis
- Syphilis
- Leprosy
- Blastomycosis
- Sarcoidosis
- Histoplasmosis
- Candidiasis
- Aspergillosis
- Actinomycosis
- Cryptococcosis
- Mucormycosis
- Rhinosporidiosis
- Lymphogranuloma

Cystic

- ABC
- Traumatic bone cyst
- Calcifying epithelial odontogenic cyst (CEOC)

Metabolic

- Hyperparathyroidism
- Paget's disease
- Fibrous dysplasia
- Cherubism
- Histiocytosis

Neoplasms

- Giant cell tumor
- Osteoblastoma
- Osteosarcoma
- Malignant lymphomas
- Wegener's granulomatosis

- Midline lethal granuloma*
- Burkitt's lymphoma
- Reticular cell sarcoma
- Pseudosarcomatous fascitis

* It is controversial issue to consider midline lethal granuloma as a malignancy as many investigators in spite of taking repeated biopsies have not found neoplastic tissue in these lesions.

Enamel Pathology**Developmental**

- Amelogenesis imperfecta
- *Dens invaginatus*
- Enameloma

Environmental pathology

- Attrition
- Abrasion
- Erosion

Enamel caries

- Pit and fissure
- Smooth surface
- Caries at cemento-enamel junction

Pigmentation

- Endogenous
- Exogenous

Enameloma**Dentin Pathology****Developmental**

- Dentinogenesis imperfecta
- Dentinal dysplasia
- Regional odontodysplasia
- Dentin hypocalcification

Dentinal caries**Neoplastic**

- Dentinoma
- Odontoma

Regressive changes

- Secondary dentin
- Dentinal sclerosis

Verrucous-papillary Lesion**Reactive lesion**

- Papillary hyperplasia of palate
- Condyloma latum
- Squamous papilloma
- Oral warts
- Oral papillomatosis

- Condyloma acuminatum
- Heck's disease

Neoplasm

- Keratoacanthoma
- Verrucous carcinoma

Unknown Etiology

- Pyostomatitis vegetans
- Verruciform xanthoma

Disorders of Tongue

Developmental

- Aglossia or hypoglossia
- Ankyloglossia
- Bifid tongue
- Lingual polyp
- Macroglossia
- Midline fistula
- Teratoma
- Median rhomboid glossitis

Infectious

- Bacterial
- Fungal and saprophytic
- Parasitic
- Viral

Cystic

- Epidermoid
- Dermoid
- Lymphoepithelial
- Mucus
- Anterior median lingual cyst
- Gastric mucosal cyst
- Parasitic cyst
- Bronchogenic cyst

Neoplastic

Benign

- Fibroma
- Granular cell myoblastoma
- Glomus tumor
- Leiomyoma
- Rhabdomyoma
- Neurofibroma
- Keratoacanthoma
- Traumatic neuroma
- Papilloma
- Pyogenic granuloma
- Adenoma
- Hemangioma
- Lymphangioma

Malignant

- Squamous cell carcinoma

- Adenocarcinoma
- Transitional cell carcinoma
- Verrucous carcinoma
- Mucoepidermoid carcinoma
- Reticular cell carcinoma
- Lymphosarcoma
- Angiosarcoma
- Kaposi's sarcoma
- Melanoma
- Rhabdomyosarcoma

Metastatic lesions from

- Kidney
- Liver
- Stomach
- Lung

Red and white lesions

- Leukoplakia
- Erythroplakia
- Lichen planus
- Oral submucous fibrosis
- Candidiasis
- Psoriasis
- Focal epithelial hyperplasia
- White sponge nevus
- Pemphigus
- Syphilitic mucus patches
- Verruca vulgaris

Neurological

- Dyskinesia—involuntary movements
- Glossodynia
- Trigeminal neuralgia
- Glossopharyngeal neuralgia
- Polyneuritis
- Neurofibromatosis
- Tongue thrusting
- Dysgeusia

Papillary changes in tongue

Atrophic

- Median rhomboid glossitis
- Geographic tongue
- Pernicious anemia
- Protein deficiency
- Lichen planus
- Oral submucous fibrosis
- Scleroderma

Hypertrophic

- White and black hairy tongue
- After antibiotic therapy
- After steroid therapy
- Hydrogen peroxide mouthwash
- Immunosuppressive drugs
- Smoking
- High fever
- Constipation
- Hyperacidity

Fissured tongue

- Congenital
- Syphilis
- Amyloidosis
- Melkersson Rosenthal syndrome
- Papillon Lefevre syndrome
- Traumatic bite

Systemic diseases manifested in tongue

- Infection—bacterial, viral and fungal
- Blood disorders
- Metabolic disorders
- Dermatological disorders
- Collagen and autoimmune disorders

Calcification of Oral Soft Tissue**Lymph nodes**

- Chronic infection (tuberculosis)
- Calcification following necrosis
- Calcification in metastatic tumor

Sialoliths

- In submandibular gland
- In minor salivary gland

Antroliths

- In maxillary antrum

Calcified ligament

- Eagle's syndrome

Osteomas

- Osteoma cutis
- Osteoma of the tongue

Calcified blood vessels

- Arteriosclerosis of facial artery
- Sturge-Weber syndrome
- Phleboliths

Myositis ossificans

- Localized
- Progressive myositis ossificans

Cysticercosis

- Calcification of worm larvae

Neoplasms

- Ossifying fibroma
- Calcifying epithelial odontogenic cyst
- Calcifying epithelial odontogenic tumor
- Cementifying fibroma
- Calcifying fibroma

Miscellaneous

- Pulp calcification
- Hypervitaminosis
- Chronic osteomyelitis
- Systemic sclerosis
- Dystrophic calcification in areas of tissue necrosis

Vascular Tissue Disorders**Arteritis**

- Polyarteritis nodosa

Midfacial granuloma syndrome

- Wegner's granulomatosis
- Stewart type of midfacial granuloma
- Giant cell arteritis
- Radiation arteritis

Vascular hamartomas

- Hemangioma
- Lymphangioma

Telangiectasia

- Hereditary hemorrhagic telangiectasia
- Radiation telangiectasia

Vascular tumor

- Hemangioendothelioma
- Hemangiopericytoma
- Kaposi sarcoma
- Angiolymphoid hyperplasia with eosinophils
- Kimura's disease

Oral Ulcerative Lesions**According to duration****Acute**

- Acute herpetic stomatitis
- Acute aphthous stomatitis
- Erythema multiforme
- Allergy
- ANUG
- Herpes zoster
- Herpangina

Chronic

- Traumatic ulcer
- Tuberculosis ulcer
- Syphilitic ulcer
- Actinomycosis ulcer
- Erosive lichen planus
- Malignant ulcer

Recurrent

- Recurrent aphthous ulcer
- Recurrent herpetic stomatitis
- Cyclic neutropenia
- Allergic reaction
- Erosive lichen planus

- Ulcer due to nutritional deficiency
- Erythema multiforme

According to Etiology

Local trauma

- Trauma due to sharp and malposed teeth
- Trauma due to appliance or restoration
- Trauma due to epileptic seizure
- Trauma during whooping cough
- Trauma from injecting needle

Chemical irritant

- Local use of chemical agents like phenol, TCA, formocresol, eugenol, phosphorus
- Local use of caustic drugs

Thermal

- Hot foods (pizza burn)
- Hot instrument
- Reverse smoking
 - Cold ice, CO₂ snow or local anesthetic or ether spray
 - Actinic radiation

Infection

Viral

- Herpes simplex
- Herpes zoster
- Chickenpox
- Smallpox
- Measles
- Rubella
- Hand foot and mouth disease
- Herpangina
- Acute lymphonodular pharyngitis
- Infectious mononucleosis
- AIDS

Bacterial

- Tuberculosis
- Syphilis
- ANUG
- Scarlet fever
- Diphtheria

Fungal infection

- Candidiasis
- Histoplasmosis
- Blastomycosis
- Mucormycosis
- Cryptococcosis

Allergy

- Local (stomatitis venenata)
- Systemic (stomatitis medicamentosa)

Neoplastic

- Squamous cell carcinoma
- Mucoepidermoid carcinoma
- Basal cell carcinoma
- Melanoma
- Malignant lymphoma

Systemic

Blood disorders

- Agranulocytosis
- Cyclic neutropenia
- Polycythemia
- Leukemia
- Aplastic anemia

Nutritional deficiency

- Vitamin B complex
- Protein deficiency
- Diabetes mellitus
- Uremia
- Metal poisoning
- Xerostomia
 - Gastric hyperacidity or peptic ulcer
- Constipation or malabsorption syndrome
- Histiocytosis-X

Disease of unknown origin

- Aphthous ulcer
- Erythema multiforme
- Epidermolysis bullosa
- Systemic lupus erythematosus
- Lichen planus
- Pemphigus
- BMMP
- Acrodermatitis enteropathica

Syndrome

- Stevens-Johnson
- Behcet's
- Reiter's

Disorders of Teeth (WHO)

Abnormalities of size and form

- Concrescence
- Fusion
- Gemination
- *Dens evaginatus*
- *Dens in dente*
- *Dens invaginatus*
- Enamel pearls
- Peg shaped
- Taurodontism
- Tuberculum paramolar
- Macrodonia
- Microdonia

Mottled teeth

- Dental fluorosis
- Mottling of enamel
- Non-fluoride enamel opacity

Disturbances in tooth formation

- Aplasia and hypoplasia of cementum

- Dilaceration of tooth
- Enamel hypoplasia (neonatal, postnatal, prenatal)
- Regional odontodysplasia (Turner tooth)

Hereditary disturbance in tooth structure

- Amelogenesis imperfecta
- Dentinogenesis imperfecta
- Odontogenesis imperfecta
- Dentinal dysplasia shell teeth

Disturbances in tooth eruption

- Dentia praecox
- Natal teeth
- Neonatal teeth
- Premature eruption of tooth
- Premature shedding of primary tooth
- Retained primary teeth

Supernumerary teeth

- Distomolar
- Fourth molar
- Mesiodens
- Paramolar
- Supplementary teeth

Anodontia

- Hypodontia
- Oligodontia

Other developmental disorders

- Embedded teeth
- Impacted teeth
- Teething syndrome
- Color change during tooth formation

Causes

Teeth Pathology

Hyperdontia

- Idiopathic
- Cleft lip and cleft palate
- Gardner's syndrome
- Cleidocranial dysplasia

Hypodontia

- Idiopathic
- Cleft lip and palate
- Hereditary hypohidrotic ectodermal dysplasia
- Incontinentia pigmenti
- Radiotherapy during childhood

Macrodontia

- Idiopathic
- Fusion
- Gemination
- Facial hemihyperplasia
- Gigantism

Microdontia

- Supernumerary teeth
- Idiopathic
- Peg shaped lateral
- Cleft lip and palate
- Hereditary hypohidrotic ectodermal dysplasia
- Radiotherapy during childhood
- Hypopituitarism

Malformed crown

- Mesiodens
- Enamel hypoplasia
- Peg shaped lateral incisor
- Turner tooth
- Talon cusp
- Amelogenesis imperfecta
- Dens evaginatus
- Dentinogenesis imperfecta
- Regional odontodysplasia
- Congenital syphilis
- Vitamin D resistant rickets
- Renal osteodystrophy
- Hypoparathyroidism
- Epidermolysis bullosa
- Radiotherapy during childhood

Enamel loss

- Caries
- Attrition
- Abrasion
- Erosion
- Amelogenesis imperfecta
- Dentinogenesis imperfecta

Extrinsic stain

- Tobacco
- Coffee, tea
- Cold drink
- Chromogenic bacteria

Intrinsic discoloration

- Aging
- Death of pulp
- Fluorosis
- Tetracycline
- Internal resorption
- Calcific metamorphoses
- Dentinogenesis imperfecta
- Amelogenesis imperfecta
- Congenital erythropoietic porphyria
- Erythroblastosis fetalis

Abnormal shaped root

- External root resorption
- Dilaceration
- Hypercementosis
- Supernumerary roots
- Concrescence

- Taurodontism
- Enamel pearl
- Benign cementoblastoma
- Radiotherapy during childhood
- Dentinogenesis imperfecta
- Dentin dysplasia type I

Enlarged pulp chamber

- Internal resorption
- Taurodontism
- Dentinogenesis imperfecta
- Regional odontodysplasia
- Vitamin D resistant rickets
- Hypophosphatasia
- Dentin dysplasia type II

Pulpal calcification

- Idiopathic pulp stone
- Secondary dentin
- Calcific metamorphosis
- Dentinogenesis imperfecta
- Dentin dysplasia type I
- Dentin dysplasia type II

Early exfoliation

- Trauma
- Aggressive periodontitis
- Immunocompromised stages
- Diabetes mellitus
- Osteomyelitis
- Cyclic or chronic neutropenia
- Langerhan's cell disease
- Dentin dysplasia type I
- Regional odontodysplasia
- Papillion-Lefevre syndrome
- Down syndrome
- Hypophosphatasia
- Scurvy

Swellings of the Palate

Traumatic

- Fracture of maxilla, alveolar process
- Hematoma
- Epulis
- Denture hyperplasia

Inflammatory

- Periapical, periodontal, gingival abscess of anterior maxillary teeth
- Osteomyelitis—acute and chronic
- Syphilis
- Tuberculosis
- Actinomycosis
- Quinsy (peritonsillar abscess)
- Infected cyst
- Mucormycosis
- Aspergillosis

- Cryptococcosis
- Toxoplasmosis

Necrosis

- Chemicals like phosphorus, arsenic
- Osteoradionecrosis
- Noma

Odontogenic cysts

- Radicular
- Residual
- Dentigerous
- Primordial
- Lateral periodontal cyst
- Gingival

Non-odontogenic cysts

- Nasopalatine or incisive canal cyst
- Globulomaxillary
- Traumatic
- Median alveolar and median palatine cyst
- Aneurysmal bone cyst
- Mucocele
- Botryoid cyst

Neoplasm

- Benign
- Malignant

Endocrine and metabolic disorders

- Fibrous dysplasia
- Paget's disease
- Hyperthyroidism
- Histiocytosis
- Caffey's disease
- Pregnancy tumor
- Leontiasis ossea

Salivary gland disorders

- Mucocele
- Tumor
- Necrotizing sialometaplasia

Developmental

- *Torus palatinus*
- Hyperplasia of palatal gland

Miscellaneous

- Impacted and supernumerary teeth
- Angioneurotic edema
- Teratoma
- Wegener's granulomatosis
- Gingival hyperplasia
- Midline lethal granuloma
- Tuberous sclerosis
- Papillomatosis in acanthosis nigricans

Swellings in the Submandibular (angle of mandible) Region**Traumatic**

- Hematoma—post-traumatic, postoperative and postextraction
- Fracture of mandible in symphysis area and body of mandible
- Denture hyperplasia
- Epulis fissuratum
- Pyogenic granuloma
- Traumatic granuloma
- *Myositis ossificans*

Cysts**Tumors****Metastasis****Inflammatory**

- Periapical, pericoronal
- Osteomyelitis
- Periostitis
- Cellulitis
- Ludwig's angina
- Tuberculosis
- Actinomycosis

Salivary Gland Dysfunction**Endocrine**

- Fibrous dysplasia
- Cherubism
- Caffey's disease

Lymphadenitis**Swelling of the Lip****Traumatic**

- From fall, assault, accidental, injury
- Lip sucking or lip biting or pipe smoking
- Trauma during epileptic and hysteric seizures
- Actinic cheilosis

Inflammatory

- Secondary to dental infection
- Carbuncle or abscess from mucus glands or hair follicle or acne
- Infected traumatic lesion
- Tuberculosis
- Leprosy
- Syphilis—chancre-gumma
- Actinomycosis
- Molluscum contagiosum
- Secondary to irradiation
- Condyloma acuminatum

Cyst

- Mucus extravasation or mucocele
- Lympho-epithelial cyst
- Epidermoid cyst
- Nasolabial cyst

Neoplasm**Allergy**

- Urticaria
- Angioneurotic edema
- Allergic macrocheilia

Developmental

- Congenital—double lip

Miscellaneous

- Amyloidosis
- Keratoacanthoma
- Papillomatosis
- Tuberous sclerosis
- Melkersson-Rosenthal syndrome
- Papillon Lefevre syndrome
- Cheilitis glandularis

Swellings in the Floor of the Mouth**Traumatic**

- Hematoma
- Fracture of alveolar process in symphysis region
- Denture granuloma
- Foreign body granuloma
- Epulis on lingual aspect of mandibular anterior teeth
- Epulis fissuratum, denture irritation hyperplasia

Inflammatory

- Abscess—periapical, gingival, periodontal, in anterior mandibular teeth
- Cellulitis—Ludwig's angina
- Infection following trauma
- Infection of Wharton's duct

Necrosis

- Chemical
- Post-irradiation

Cysts**Odontogenic**

- Radicular
- Dentigerous
- Gingival
- Residual
- Keratocyst
- Primordial

Non-odontogenic

- Traumatic
- Ranula
- Mucocele
- Sublingual dermoid cyst
- Lympho-epithelial cyst
- Cystic hygroma

Syndrome

- Sturge-Weber syndrome

Neoplasms**Benign epithelial tumors**

- Papilloma
- Adenoma
- Oncocytoma
- Oxyphilic adenoma

Malignant epithelial tumors

- Squamous cell carcinoma
- Malignant melanoma
- Mucoepidermoid carcinoma
- Adenocarcinoma
- Transitional cell carcinoma

Benign connective tissue tumors

- Hemangioma
- Lymphangioma
- Neurofibroma
- Leiomyoma
- Rhabdomyoma
- Fibroma
- Lipoma
- Granular cell myoblastoma

Malignant mesenchymal and other tumors

- Fibrosarcoma
- Hemangioendothelioma
- Reticular cell sarcoma
- Rhabdomyosarcoma
- Malignant lymphoma

Salivary gland swelling

- Sialadenitis
- Sialolithiasis
- Sjögren's syndrome
- Mikulicz's disease
- Salivary gland tumors
- Cysts of salivary gland

Allergy

- Angioneurotic edema

Swelling of Neck**Lateral neck swelling**

- Lymphadenitis
- Metastatic carcinoma to lymph nodes
- Lymphoma
- Parotid lesion
- Metabolic diseases
- Carotid body tumor
- Epidermoid cyst
- Cystic hygroma

Midline neck swelling

- Thyroglossal duct cyst
- Thyroid tumor
- Dermoid cyst

Extraoral Discharging Sinus**Developmental**

- Salivary fistula
- First or second branchial arch sinus or fistula
- Congenital lip pit

Inflammatory

- Periapical, periodontal and dentoalveolar abscess pointing extraorally
- Chronic osteomyelitis
- Syphilis
- Tuberculosis of jaw bone
- Actinomycosis
- Osteoradionecrosis
- Salivary fistula from suppurative sialadenitis
- Infected lymph node
- Infected cyst
- Infection secondary to trauma, injury
- Infection of various facial space
- Oronasal fistula

Neoplastic

- Intraoral malignancy with secondary growth
- Salivary gland malignancy

Perforations of the Palate**Congenital**

- Cleft palate

Inflammatory

- Osteomyelitis
- Syphilitic gumma
- Tuberculosis
- Infection of traumatic and chronic wound
- Mucormycosis
- Toxoplasmosis
- Blastomycosis
- Leprosy
- Wegener's granulomatosis

Traumatic

- Fracture of maxilla
- Use of suction disc for dentures
- Pizza burn, thermal burn

Neoplasm

- Squamous cell carcinoma
- Mucoepidermoid carcinoma
- Adenocystic carcinoma
- Osteosarcoma
- Lymphoma
- Midline lethal granuloma

Glossodynia**Local causes**

- Traumatic injury

- Median rhomboid glossitis
- Geographic tongue
- Lichen planus
- Oral submucous fibrosis
- Candidiasis
- Denture irritation
- Allergic reaction to denture material
- Malignant lesions
- Ulcerative and erosive lesions on tongue due to infection, trauma, allergy and systemic disease
- Xerostomia
- Trauma to lingual nerve
- Fusospirochetal infection
- TMJ dysfunction
- Chronic mouth breathing

Systemic diseases

- Diabetes
- Neuralgia—lingual, glossopharyngeal and facial
- Neuritis
- Nutritional deficiency
- Allergy
- Anemia
- Sjögren's syndrome
- Chronic gastritis
- Hypothyroidism
- Mercurialism
- AIDS

Psychological

- Emotional stress and strain
- Depression
- Neurosis
- Schizophrenia
- Mood disorders

Halitosis

Physiological

- *During infancy*—sweet odor
- *During development of dentition*—pungent odor
- Menstruation
- *Food habits*—alcohol, garlic, lemon, other aromatic beverages or food substances
- *Old age*—due to metabolic changes in periodontal tissue
- Hunger breath

Pathological

Local oral conditions

- Retention of food around the teeth (interdental area, flaps)
- Gingival and periodontal disease
- Deep carious lesion and necrotic pulp
- Coating of tongue
- Mouth breathing
- Denture and orthodontic appliance
- Oral soft tissue lesion
- Stomatitis
- Glossitis

- Pharyngitis
- *Habits*—smoking, tobacco and catechu

Systemic conditions

- *Odor from nasopharynx*—due to adenoma, rhinitis, sinusitis, postnasal drip, tonsillitis, tonsillar abscess, pharyngitis
- *From lung and bronchi*—bronchitis, lung abscess, pulmonary tuberculosis, lung malignancy
- *Diseases of gastrointestinal tract*—hyperacidity, gastritis, peptic ulcer, malignancy
- *Metabolic disorders*—diabetes, uremia and liver disease
- *Blood disorders*—anemia, agranulocytosis, leukemia, thrombocytopenia
- Salivary gland dysfunctions causing xerostomia
- *Dehydration states*—diarrhea, vomiting, diabetes insipidus,
- *Drug*—isosorbide dinitrate, iodine derivatives, diuretics, phenothiazines, immunosuppressive drugs and dimethyl sulfoxide

Enlargement of Cervical Lymph Nodes

Inflammation and Infection

Bacterial infections

- Specific bacterial infection
 - Syphilis
 - Tuberculosis
- Non-specific
 - Periodontal disease
 - Pericoronitis
 - Periapical infections

Viral infections

- Infection caused by herpes simplex
- Human immunodeficiency virus
- Cat scratch disease
- Infectious mononucleosis

Fungal infection

- Histoplasmosis
- Oral candidiasis

Parasitic infection

- Rickettsial infections

Allergic conditions

- Serum sickness

Primary neoplasm

- Lymphoma

Metastasis tumors

- Oral squamous cell carcinoma
- Metastasis carcinoma from the breast

Miscellaneous conditions

- Non-tender lymphoid hyperplasia
- Collagen disease
- Sarcoidosis
- Leukemia

Trismus**Traumatic****Intra-articular**

- Trauma to growth center or developing condyle
- Monoarticular arthritis like spasm or subluxation (due to chronic malocclusion)
- Trauma to articular disc
- Trauma to capsular or temporomandibular ligament
- Fracture with or without dislocation and displacement of condylar head
- Ankylosis
- Foreign body in the joint area

Extra-articular

- Trauma to muscle
- Trauma during extraction
- Faulty mandibular block
- Fracture of coronoid process
- Fracture of zygomatic arch
- Fracture of styloid process
- Misplaced fixative or augmentative device

Inflammatory**Intra-articular**

- Pyogenic infection
- Rheumatic fever
- Rheumatoid arthritis
- Gout
- Tuberculosis
- Syphilis
- Actinomycosis
- Osteomyelitis
- Synovitis

Extra-articular

- Pericoronitis
- Acute and chronic osteomyelitis
- Acute and chronic dentoalveolar abscess

- Cancrum oris or noma
- Abscess in various fascial spaces like infratemporal, lateral pharyngeal, submasseteric, and pterygomandibular spaces
- Phosphorus necrosis
- Quinsy (peritonsillar abscess)
- Infection of ear or otitis media
- Mumps
- Acute generalized stomatitis
- Angular cheilitis
- Pharyngitis

Congenital and developmental disorders

- Unilateral or bilateral condylar hyperplasia
- Unilateral or bilateral exostosis of coronoid
- Hyperplasia of coronoid process and zygomatic arch
- Fusion of coronoid process with zygomatic arch
- Absence of coronoid process

Neoplastic**Intra-articular****Benign**

- Osteoma
- Chondroma
- Osteochondroma
- Myxoma
- Benign giant cell tumor
- Trotter's syndrome

Malignant

- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma
- Metastatic carcinoma

Extra-articular

- Benign and malignant tumors of mandible and maxilla mainly in premolar and molar area

Summary table of the effects of different inflammatory processes on the periapical tissues and the resultant radiographic appearances

Stage of inflammation	Underlying inflammatory changes	Radiographic appearance
Initial acute inflammation	Inflammatory exudate accumulates in the apical periodontal ligament space—acute apical periodontitis	Widening of the radiolucent line of periodontal ligament space or no apparent changes evident
Initial spread of inflammation	Resorption and destruction of the apical bony socket—periapical abscess	Loss of radiopaque line of lamina dura at the apex
Further spread of inflammation	Further resorption and destruction of the apical alveolar bone	Area of bone loss at the tooth apex
Initial low-grade chronic inflammation	Minimal destruction of the apical bone. The body's defense systems lay down dense bone in the apical region	No apparent bone destruction but dense sclerotic bone evident around the tooth apex (sclerosing osteitis)
Latter stages of chronic inflammation	Apical bone is resorbed and destroyed and dense bone is laid down around the area of resorption—periapical granuloma or radicular cyst	Circumscribed well defined radiolucent area of bone loss at the apex, surrounded by dense sclerotic bone

- Malignant tumors of nasopharynx
- Malignant tumors of maxillary sinus
- Parotid tumors

Cystic

Extra-articular

- All large cystic lesion affecting mandible, ramus and posterior part of maxilla

Systemic

- Tetanus
- Tetany
- Fibrous dysplasia
- Paget's disease
- Acromegaly

Miscellaneous

- Epilepsy and hysteria
- Hydrophobia
- Sudden physical, chemical and electrical shock
- Hemorrhage in medulla oblongata
- Brain tumor
- Scleroderma
- Strychnine poisoning
- Myelofibrosis
- Amylotrophic lateral sclerosis
- Oral submucous fibrosis
- Osteoradionecrosis
- Myositis ossificans
- Disuse atrophy
- Myofascial pain dysfunction syndrome (MPDS)
- Extensive burns of face with keloid and scar formation
- Dystrophic epidermolysis bullosa

Hemorrhage

Local causes

- Post-extraction, post-surgical and post-traumatic
- Infections—viral, bacterial, fungal, parasitic and spirochete
- Oral ulcerative lesions—stomatitis, glossitis
- Oral exophytic soft tissue lesions—pyogenic granuloma, pregnancy tumor
- Local irritants leading to gingivitis and periodontitis
- Rupture of blood containing bulla
- Congenital hamartomas—hemangioma, hereditary hemorrhagic telangiectasia, arteriovenous malformation

Hemorrhage due to platelet disorders

- Thrombocytopenia
- Thrombocytosis
- Thrombasthenia
- Glanzmann's disease
- Aldrich syndrome

Hemorrhage due to coagulation diseases

- Hemophilia
- Christmas disease

- von Willebrand's disease
- Deficiency of Stuart factor
- Multiple myeloma
- Systemic lupus erythematosus
- Diffuse intravascular coagulation
- Macroglobulinemia

Hemorrhage due to systemic diseases

- Scurvy
- Diabetes mellitus
- Septic embolism in bacterial endocarditis
- Meningococemia
- Systemic viral infection
- Allergy
- Anti-coagulant therapy
- Graft versus host reaction
- Sturge-Weber syndrome

Angular cheilitis

- Infections
 - Candidiasis
 - Herpes labialis
- Unhygienic appliances
- Malabsorption syndrome
- Diabetes
- Change in vertical dimension
- Xerostomia
- Nutritional deficiency
 - Anemia
 - Vitamin deficiency
 - Protein deficiency
 - Plummer-Vinson syndrome
- Split papule of syphilis
- Allergic reaction to lipstick
- Cold sore

Sialorrhea

- Acute inflammation of oral mucosa
- Fracture of jaw bone
- During eruption of teeth in infants
- Mental retardation
- Neurosis
- Psychosis
- Parkinsonism
- Schizophrenia
- Epilepsy
- Acrodynia (mercury poisoning)
- Rabies
- Familial dystonia
- Drugs like sialogogue

Xerostomia

Factor affecting salivary center

- Emotional disturbance like stress, strain
- Depression

- Hysteria
- Neurosis

Factor affecting autonomous nervous system

- Encephalitis
- Brain tumor
- Neurological operation

Factor affecting salivary gland

- Developmental
- Inflammatory
- Tumor of salivary gland
- Sialolithiasis
- Sjögren's syndrome
- Mikulicz's disease
- Atrophy of gland

Alteration in fluid and electrolyte balance

- Dehydration
- Diarrhea
- Vomiting
- Diuresis
- Diabetes insipidus
- Congestive cardiac failure
- Ascites
- Liver cirrhosis

Drugs

- Anticholinergic
- Antidepressants
- Sympathomimetics
- Opium derivatives
- Antipsychotic
- Antihistamine
- Diuretics
- Sedatives
- Digitalis
- Steroids
- Chemotherapeutic agents

Miscellaneous

- Malnutrition
- Nutritional and vitamin deficiency
- Radiation
- Toxemia
- Habits (smoking, betel nut chewing)
- Chronic alcoholism

Saliva Drooling at Corner of Mouth

- Oral infection, throat infection
- Jaw fracture
- Psychosis
- Schizophrenia
- Rabies
- Acrodynia
- During eruption of teeth in infant

Discoloration of the Teeth

Extrinsic

- Habits—tobacco, catechu
- Chromogenic bacteria
- Poor oral hygiene
- Oral drugs
- Iatrogenic
- Tattoo made by patient
- Chlorhexidine mouthwash
- Medicaments like AgNO₃, iodine, iron

Intrinsic

- Erythroblastosis fetalis due to Rh incompatibility
- Neonatal jaundice
- Congenital porphyria
- Tetracycline therapy during formation of teeth
- Cystic fibrosis
- Osteogenesis imperfecta
- Dentinogenesis imperfecta
- Amelogenesis imperfecta
- Fluorosis
- Enamel opacities
- Non-vital teeth or internal resorption
- Congenital heart disease
- Acute exanthematous disease
- Turner's teeth

Macroglossia

Congenital or developmental

- Mongolism
- Lingual thyroid or polyp

Inflammatory

- Syphilis
- Amoebic dysentery
- Ludwig's angina
- Pneumonia
- Typhoid
- Tuberculosis
- Blastomycosis
- Infected wound
- Actinomycosis

Neoplasms

- Hemangioma (diffuse type)
- Neurofibromatosis
- Lymphangioma (diffuse type)

Systemic

- Pellagra
- Down's syndrome
- Myxedema
- Acromegaly
- Amyloidosis
- Uremia
- Gardner's syndrome
- Diabetes

- Scurvy
- Dyskinesia chorea
- Melkersson-Rosenthal syndrome
- Sturge-Weber syndrome
- Hurler's syndrome
- Beckwith's hypoglycemia syndrome
- Tuberous sclerosis

Orofacial pain

Extra-cranial causes

Dental and oral

- Dentinal hypersensitivity
- Pain from disorders of pulp
 - Hyperemia
 - Acute pulpitis
 - Chronic pulpitis
- Pain from disorders of periodontium
- Mucogingival pain
 - Stomatitis
 - Gingivitis
 - Glossitis
 - Glossodynia
- Osseous and periosteal pain
 - Dry socket
 - Periostitis
 - Osteomyelitis
- From disorders of ear and eye
- Enlargement of salivary glands

Pain from paranasal sinuses

- Sinusitis
- Tumors of the sinus

Musculoskeletal

- Myofacial pain
- Temporomandibular joint arthropathy
- Cervical spine disorders
- Trotter's syndrome
- Eagle's syndrome

Intra-cranial causes

- Traction disorders of pain sensory structure of the brain
- Neoplasms, aneurysms, hematoma, hemorrhage or edema
- Displacement of great venous sinus
- Distortion and dilation of intracranial vessels

Vascular pain

- Migraine headache
- Cluster headache
- Tension headache
- Temporal headache
- Carotodynia
- Angina pectoris or myocardial infarction

Neurogenic pain

Paroxysmal

- Trigeminal neuralgia
- Glossopharyngeal neuralgia
- Geniculate neuralgia

Differentiation pain syndrome

- Atypical odontalgia
- Traumatic neuroma
- Neuritis
- Reiter's sympathetic dystrophy

Psychogenic pain

- Anxiety and depression
- Delusional or hallucination pain
- Hysterical or hypochondriac pain

Classification by Bells

Somatic

- Superficial—mucogingival
- Deep visceral—musculoskeletal

Neurogenic

- Neuropathy—neuralgia and neuritis
- Deafferentation syndrome

Psychogenic

- Conversion hysteria
- Delusional pain

Sloughing Pseudomembranous Necrotic White Lesion

- Plaque
- Traumatic ulcer
- Diffuse gangrenous stomatitis
- Diphtheria
- Noma
- Candida endocrinopathy syndrome
- Chemical burns
- ANUG
- Candidiasis
- Eosinophilic granuloma
- Superficial abscess
- Syphilitic chancre and mucous patches

Osteoporosis

- Osteogenesis imperfecta
- Post-menopausal osteoporosis
- Hyperthyroidism
- Diabetes
- Old age
- Gonadal osteoporosis
- Idiopathic osteoporosis
- Renal acidosis
- Oxalosis
- Acromegaly
- Adaptation syndrome
- Hypervitaminosis D
- Hypovitaminosis C

Depapillation of Tongue

Congenital cause

- Familial dystonia
- Epidermolysis bullosa
- Dyskeratosis congenita
- Endocrine candidiasis

Developmental

- Geographic tongue
- Median rhomboidal glossitis
- Central papillary atrophy

Chronic trauma**Nutritional and hematological abnormalities**

- Pellagra
- Riboflavin
- Conditional deficiency
- Plummer-Vinson syndrome

Medication

- Antibiotic
- Anticholinergic agent
- Cancer chemotherapeutic agent

Peripheral vascular disease**Chronic candidiasis****Tumor**

- Squamous cell carcinoma
- Epidermoid carcinoma

Miscellaneous

- Diabetes mellitus
- Oral submucous fibrosis

Atrophic Lesion of Oral Mucosa of Tongue**Developmental**

- Benign migratory glossitis
- Median rhomboidal glossitis

Systemic condition

- Endocrine—diabetes mellitus
- Vitamins like B₂, B₅, B₆, B₉ and B₁₂ deficiency
- Blood disorders—pernicious anemia and iron deficiency anemia

Infection

- Syphilis
- Chronic candidiasis

Oral submucous fibrosis**Mucocutaneous**

- Atrophic lichen planus
- Discoid lupus erythematosus
- Dyskeratosis congenita
- Scleroderma
- Xerostomia

Retarded Eruption of Teeth**Local causes**

- Loss of space

- Abnormal crypt position
- Overcrowding
- Additional teeth
- Retention of deciduous predecessor
- Dentigerous and eruption cyst
- Hereditary gingival fibromatosis

Systemic causes

- Congenital hypopituitarism
- Congenital hypothyroidism
- Down syndrome
- Cleidocranial dysplasia
- After radiation
- Rickets

Loosening or Early Loss of Teeth**Local causes**

- Inflammatory periodontal disease
- Trauma
- Juvenile periodontitis

Systemic causes

- Down syndrome
- Diabetes mellitus
- Neutropenia
- Hypophosphatasia
- Papillon lefevre syndrome
- Ehlers-Danlos syndrome
- AIDS related disorders

Others

- Acrodynia
- Neoplasm
- Eosinophilic granuloma

Malformed Teeth

- Local infection or trauma
- Radiotherapy
- Congenital syphilis
- Down syndrome
- Ectodermal dysplasia
- Rickets

Hairy Tongue

- Dehydration
- Debilitated patient
- Long illness
- Painful oral condition restricted tongue movement
- Use of local and systemic medication like systemic antibiotic, topical H₂O₂ and perborate

Causes that Cause Red Lesions**Marked increased in hemoglobin concentration of articulating blood**

- Polycythemia

Vascular dilation from**Inflammation (erythema)**

- Mechanical trauma (cheek biting, ill fitting denture)
- Thermal trauma (hot food)
- Chemical trauma
- Infection (cellulitis and Ludwig angina)
- Allergy or autoimmune disease
- Ulcer with inflamed rim (recurrent herpetic ulcer)

Congenital defect

- Hemangioma

Extravasation of blood (trauma or homeostatic disease)**Atrophy or thinning of mucosa****Leukopenia****Infections****Bacterial**

- Typhoid
- Paratyphoid fever
- Brucellosis
- Tularemia (early)

Viral and rickettsial

- Influenza
- Measles
- Rubella
- Chickenpox
- Infectious hepatitis
- Colorado tick fever
- Dengue
- Yellow fever

Protozoal

- Malaria
- Relapsing fever
- Kala azar

Any overwhelming infection

- Miliary tuberculosis
- Septicemia

Hemopoietic disorders

- Gaucher's disease
- Pernicious anemia
- Aplastic anemia
- Chronic hypochromic anemia
- Aleukemic leukemia
- Agranulocytosis

Chemical agents**Agents commonly producing leukopenia in all patient if given in sufficient dose**

- Mustards (sulfur and nitrogen mustards)
- Urethane
- Busulfan
- Benzene
- Antimetabolites

Agents occasionally associated with leukopenia apparently as a result of individual sensitivity

- Analgesics, sedative and anti-inflammatory
- Antithyroid drug
- Anticonvulsant
- Sulfonamides
- Antihistamine
- Antimicrobial agents
- Tranquilizers

Physical agents

- X-ray radiation and radioactive substance

Anaphylactic shock and early stages reaction of foreign protein**Disease of unknown etiology**

- Liver cirrhosis
- Disseminated erythematosis
- Cyclic neutropenia

Basophilia**Blood disorders**

- Chronic myelocytic leukemia
- Chronic anemia
- Hodgkin's disease

Splenectomy**Infection**

- Chronic inflammation of accessory tissue
- Smallpox
- Chickenpox

Myxedema**After injection of foreign proteins****Some cases of nephrosis****Neutrophilia****Acute infection**

- Coccal
- Bacilli
- Fungi
- Spirochetes
- Virus
- Rheumatic fever
- Diphtheria
- Smallpox

Inflammatory

- Coronary thrombosis
- Gout
- Collagen vascular disease
- Burns
- Hypersensitivity reaction

Intoxication

- Uremia

- Diabetes acidosis
- Poisoning by chemical and drugs like lead, mercury, digitalis, insect venoms, black widow spider

Acute hemorrhage*Acute hemolysis***Malignant tumor of**

- Gastrointestinal tract
- Liver
- Bone marrow

Blood disorders

- Myelocytic leukemia
- Polycythemia
- Myelofibrosis
- Myeloid metaplasia
- Chronic idiopathic neutropenia
- Hereditary neutrophilia

Miscellaneous

- Physiologic in the newborn
- During labor
- After repeated vomiting
- Convulsion
- Paroxysmal tachycardia
- After epinephrine injection

Eosinophilia*Allergic*

- Bronchial asthma
- Urticaria
- Angioneurotic edema
- Hay fever
- Allergic rhinitis
- Drug sensitivity

Skin disease

- Pemphigus
- Dermatitis herpetiformis
- Bullous pemphigoid

Parasitic infection

- Trichinosis
- Echinococcosis disease

Blood disorders

- Chronic myelocytic leukemia
- Polycythemia vera
- Hodgkin's disease
- Pernicious anemia

Infection

- Scarlet fever
- Chorea
- Erythema multiforme

Malignant disease of any type

Following irradiation

Miscellaneous

- Pulmonary infiltration with eosinophilia
- Tropical eosinophilia
- Polyarteritis nodosa
- Rheumatoid arthritis
- Sarcoidosis
- Certain poison

Inherited**Idiopathic****Lymphocytosis****Acute infection**

- Infectious mononucleosis
- Acute infectious lymphocytosis
- Infectious hepatitis

Chronic infection

- Tuberculosis
- Secondary and congenital syphilis
- Undulant fever

Lymphocytic leukemia**Lymphosarcoma****Heavy chain disease****Hemopoietic disorders**

- Neutropenia
- Exanthema

Monocytosis**Bacterial infection**

- Tuberculosis
- Subacute bacterial endocarditis
- Syphilis
- Brucellosis
- Typhoid

Protozoan and rickettsial infection

- Malaria
- Rocky Mountain spotted fever
- Typhus
- Kala azar
- Trypanosomiasis
- Oriental sore

Blood disorders

- Lymphoma
- Leukemia
- Hodgkin's disease
- Multiple myeloma

Lipid storage disease

- Gaucher disease

Malignant neoplasm

- Carcinoma of ovary, breast and stomach

Collagen vascular disease

- Lupus erythematosus
- Rheumatoid arthritis

Granulomatous disease

- Sarcoidosis
- Ulcerative colitis
- Regional arteritis

Chronic high dose steroid therapy**Peripheral Plasmocytosis****Infection****Viral**

- Rubella
- Rubeola
- Varicella
- Infectious mononucleosis

Bacterial

- Streptococcal
- Diplococcal
- Syphilis
- Tuberculosis

Protozoal

- Malaria
- Trichinosis

Serum Sickness**Drugs**

- Penicillin
- Sulfisoxazole

Antitoxins

- Equine tetanus
- Equine diphtheria

Neoplasm**Hematological**

- Plasma cell leukemia
- Chronic lymphocytic leukemia

Nonhematological

- Breast
- Prostate

Miscellaneous

- Transfusion
- Hyperimmunization
- Trauma

Periapical Radiolucency

- Acute apical periodontitis
- Periapical abscess
- Periapical granuloma
- Periapical cyst
- Dentigerous cyst

- Periapical scar
- Giant cell granuloma
- Benign and malignant tumor including secondary metastatic deposits
- Lymphoreticular tumors of bone
- Langerhan's cell disease
- Periapical cemental dysplasia
- Surgical defect
- Osteomyelitis

Monolocular and Multilocular Radiolucency**Monolocular lesion**

- Radicular cyst
- Residual cyst
- Dentigerous cyst
- Lateral periodontal cyst
- Nasopalatine duct cyst
- Simple bone cyst
- Calcifying epithelial odontogenic tumor
- Adenomatoid odontogenic tumor
- Primary bone tumors
- Secondary metastasis tumor
- Multiple myeloma
- Eosinophilic granuloma
- Fibro-cemento-osseous lesion
- Stafne's bone cavity

Multilocular lesion

- Odontogenic keratocyst
- Ameloblastoma
- Ameloblastic fibroma
- Ameloblastic fibro-odontoma
- Odontogenic myxoma
- Central giant cell granuloma
- Brown tumor
- Cherubism
- Aneurysmal bone cyst
- Metastatic tumor of the jaw
- CEOT
- Fibrous dysplasia
- Burkitt's lymphoma
- Squamous odontogenic tumor

Pericoronal Radiolucency

- Dentigerous cyst
- Ameloblastoma
- Calcifying odontogenic cyst
- Adenomatoid odontogenic tumor
- Pericoronal space
- Unicystic ameloblastoma
- Ameloblastic fibroma
- Envelopmental primordial cyst

Inter-radicular Radiolucency

- Primary tooth crypt
- Mental foramina
- Nutrient canal
- Bony periodontal pocket

- Malignancy
- Lateral periodontal cyst
- Median mandibular cyst
- Paradental cyst
- Furcation involvement
- Lateral radicular cyst
- Traumatic bone cyst
- Odontogenic tumor
- Globulomaxillary tumor
- Incisive canal cyst
- Histiocytosis-X
- Osteoradionecrosis

Solitary Cyst-like Radiolucency not Necessary Contacting Teeth

- Primordial cyst
- Traumatic cyst
- Residual cyst
- Odontogenic keratocyst
- Aneurysmal bone cyst
- Cementoma
- Central fibroma
- Ameloblastoma
- Giant cell granuloma
- Midpalatal cyst
- Cementifying ossifying fibroma
- Benign non-odontogenic tumor
- Central squamous cell carcinoma
- Myxoma

Multiple Separate Well-defined Radiolucency

- Multiple myeloma
- Basal cell nevus syndrome
- Multiple cyst or granuloma
- Histiocytosis-X
- Cherubism
- Nodular central masses
- Neurofibromatosis
- Hurler's and Hunter's syndrome

Generalized Rarefaction of Jaw Bones

- Hyperparathyroidism
- Osteoporosis
- Osteogenic imperfecta
- Hypervitaminosis D
- Diabetes
- Osteomalacia
- Leukemia
- Paget's disease
- Multiple myeloma
- Lymphosarcoma

Mixed Radiolucent—Radiopaque Lesion Associated with Teeth

- Calcifying crown of developing teeth
- Tooth root with rarefying osteitis
- Rarefying and condensing osteitis
- Cementifying and ossifying fibroma
- Periapical cementoma—intermediated stage
- Foreign bodies

- Odontoma
- Osteomyelitis chronic

Pericoronal Mixed Lesion

- Odontoma—intermediate stage
- AOT
- Keratinizing and calcifying odontogenic cyst
- CEOT
- Odontogenic fibroma
- Eruption sequestrum
- Cystic odontoma
- Ameloblastic fibroma

Mixed Radiolucent-radiopaque Lesion not Necessary Contacting Teeth

- Chronic osteomyelitis
- Osteoradionecrosis
- Fibrous dysplasia
- Paget's disease—intermediate stage
- Central hemangioma
- Osteoid osteoma
- Cementifying and ossifying fibroma
- Osteogenic sarcoma
- Chondroma and chondrosarcoma
- Ossifying subperiosteal hematoma
- Calcifying epithelial odontogenic tumor

Variable Radiopacities

Abnormalities of the teeth

- Unerupted and misplaced teeth including supernumeraries
- *Odontoma*—compound and complex
- Root remnants
- Hypercementosis

Condition affecting the bone

Developmental

- *Exostosis including tori*—mandibular or palatal

Inflammatory

- *Low grade chronic infection*—sclerosing osteitis
- *Osteomyelitis*—sequestra involucrum formation

Tumors odontogenic (late stages)

- Calcifying epithelial odontogenic tumor
- Adenomatoid odontogenic tumor
- Calcifying odontogenic cyst

Non-odontogenic tumor

- *Benign*—osteoma, chondroma
- *Malignant*—osteosarcoma, osteogenic secondary metastases

Fibro-cemento-osseous

- Fibrous dysplasia
- Periapical cemental dysplasia
- Gigantiform cementoma
- Benign cementoblastoma
- Cemento-ossifying fibroma

Other

- Paget's disease
- Osteopetrosis

Superimposed soft tissue calcifications

- Salivary calculi
- Calcified lymph node
- Calcified vessels
- Phleboliths
- Calcified ACE scars

Foreign bodies

- Intrabony
- Within the soft tissue
- On or overlying the skin

Periapical radiopacities**True**

- Condensing osteitis
- Periapical idiopathic osteosclerosis
- Mature periapical cementoma
- Foreign bodies
- Hypercementosis

Projected

- Sialoliths
- Phleboliths
- Arterial calcification
- Retained root tips
- Exostosis tori and periapical osteoma
- Calcified lymph nodes

Multiple Separate Radiopacities

- Multiple chondroma
- Sickle cell sclerosis
- All solitary radiopacities not necessary contacting teeth

Solitary Radiopacities not Necessary Containing Teeth**True**

- Tori, exostosis and periapical osteoma
- Unerupted, impacted and supernumerary teeth
- Retained root
- Idiopathic osteosclerosis
- Cementifying and ossifying fibroma
- Chondroma
- Chondrosarcoma
- Mature osteoblastoma
- Osteogenic sarcoma
- Fibrous dysplasia
- Mature cementoma
- Sclerosing osteomyelitis and diffuse sclerosing osteomyelitis
- Proliferative periostitis

Projected

- Anatomic radiopacities
- Foreign bodies
- Pathologic soft tissue
- Arterial calcification
- Rhinoliths and antroliths

Generalized Radiopacities

- Gardener's syndrome
- Fluorosis
- Caffey's disease
- Adenomatoid odontogenic tumor
- Metastatic carcinoma of prostate
- von Buchan disease
- Osteopetrosis

Thickened Periodontal Ligament Space

- Periapical abscess
- Current orthodontic therapy
- Increase occlusal friction
- Systemic sclerosis
- Sarcoma infiltration
- Carcinoma infiltration

Loss of Lamina Dura

- Periapical infections
- Fibrous dysplasia
- Paget's disease
- Osteoporosis
- Normal anatomical variation
- Hyperparathyroidism
- Leukemia
- Cushing syndrome
- Hypophosphatasia
- Osteomalacia
- Multiple myeloma

Different Type of Periosteal Reaction and their Cause**Regular type****Parallel type**

- Onion peel appearance
- Eosinophilic granuloma
- Chronic osteomyelitis
- Codman's triangle
- Osteosarcoma

Radiating type

- Sunray appearance
- Osteosarcoma
- Hair on end appearance
- Paget's disease
- Central hemangioma

Irregular type

- Osteomyelitis
- Fracture
- Osteogenic malignancy
- Subperiosteal bleeding
- Leukemia
- Histiocytosis
- Arthritis
- Rheumatism

Typical Features of the Lesion*Typical clinical feature*

<i>Name of disease</i>	<i>Typical feature</i>
Ameloblastoma	Egg shell cracking
Pyogenic granuloma	Hour-glass appearance
Hodgkin's disease	Bimodal age incidence peak
Psoriasis	Auspitz's sign
Lichen planus	Wickham's striae
Pemphigus	Nikolsky's sign
Scarlet fever	Strawberry and Raspberry tongue
Diphtheria	Diphtheritic membrane
Wegener's granulomatous	Strawberry gingivitis
Tetany	Risus sardonicus
Behcet's syndrome	Pathergy test positive
Measles	Koplik's spot
Mild restricted muscular dystrophy	Tapir-lips
Leukokeratosis nicotina glossi	Golf ball appearance
Bell's palsy	Masklike or expressionless appearance
Rickets	Rachitic rosary, pigeon breast
Pseudoxanthoma elasticum	Hound dog

Typical radiological feature

<i>Typical radiological feature</i>	<i>Name of disease</i>
Soap bubble appearance	Ameloblastoma, aneurysmal bone cyst, central hemangioma
Honeycomb pattern	Calcifying epithelial odontogenic cyst, odontogenic myxoma, hemangioma, central giant cell granuloma
Driven snow appearance	Calcifying epithelial odontogenic cyst
Sun burst appearance	Hemangioma, osteosarcoma
Sunray appearance	Osteosarcoma, hemangioma, osteoblastoma
Hair on end appearance	Sickle cell anemia, thalassemia
Tennis racket	Odontogenic myxoma
Moth eaten	Early stage of osteosarcoma, squamous cell carcinoma, osteomyelitis, osteoradionecrosis, leukemia, malignant lymphoma
Teeth standing in space or floating teeth	Histiocytosis-X, severe periodontitis, malignant lymphoma
Ground glass	Fibrous dysplasia, Paget's disease, hyperparathyroidism, ossifying fibroma
Orange peel	Fibrous dysplasia
Cotton wool	Paget's disease
Downward bowing	Cemento-ossifying fibroma
Mass of coral	Calcified lymph node
Antral halo	Acute sinusitis

<i>Typical radiological feature</i>	<i>Name of disease</i>
Hanging drop appearance in maxillary sinus	Blow out orbital fracture
Ely's cyst	Osteoarthritis
Pepper pot skull	Hyperparathyroidism
Step ladder pattern	Sickle cell anemia
Tree in winter appearance	Normal sialographic appearance of parotid gland
Bush in winter appearance	Normal sialographic appearance of submandibular gland
Sausage link appearance	Sialographic appearance of sialodochitis (ductal inflammation/infection)
Snowstorm appearance	Sialographic appearance of Sjögren's syndrome
Sialectasis	Sialographic appearance of sialadenitis
Ball in hand appearance	Sialographic appearance of intrinsic benign tumor
Codman's triangle	Osteogenic sarcoma
Balloon like appearance	Follicular cyst
Candlestick appearance	Pyknodysostosis and progressive systemic sclerosis
Chalk like appearance	Osteopetrosis, pyknodysostosis, hyperparathyroidism
Cherry blossom pattern	Sialographic appearance of Sjögren's syndrome
Eggshell appearance	Ameloblastoma, multilocular cyst
Filling defect	Salivary gland tumor
Mottled appearance	Fibrous dysplasia, ossifying fibroma
Onion peel (skin) appearance	Chronic osteomyelitis, eosinophilic granuloma, Ewing's sarcoma
Permeated type	Carcinoma of gingiva, squamous cell carcinoma of maxilla
Pear shaped appearance	Globulomaxillary cyst
Pencil line appearance	Ameloblastoma, traumatic bone cyst and CEOC
Pressure type appearance	Squamous cell carcinoma of the gingiva
Punched out appearance	Multiple myeloma
Sand like appearance	Adenoameloblastoma, CEOC and CEOT
Salt and paper appearance	Hyperparathyroidism, ABC and giant cell granuloma
Scalloping pattern (margins)	Dentigerous cyst, traumatic bone cyst, ABC and giant cell tumor
Spiked root	Malignant histiocytoma, Burkitt's lymphoma
Beaten silver	Craniofacial dysostosis (Crouzon' disease)
Sharpened pencil or mouth piece of flute	Rheumatoid arthritis of TMJ
Thumb print	Fibrous dysplasia

Typical histological features

<i>Typical histological features</i>	<i>Name of disease</i>
Starry sky	Burkitt's lymphoma, infective mononucleosis, polycythemia
Rushton bodies	Dentigerous cyst
Reed-Sternberg cell	Hodgkin's disease
Dropping off effect	Junctional nevus
Saw tooth appearance	Lichen planus
Antoni type A and type B tissue	Neurilemmoma
Honeycomb or Swiss cheese pattern	Adenoid cystic carcinoma of salivary gland
Picket fence or tombstone	Primordial cyst
Liesegang ring	Calcifying epithelial odontogenic tumor
Safetypin appearance	Granuloma inguinale
Lipschutz bodies	Herpes simplex infection
Anitschkow cell	Aphthous ulcer, sickle cell anemia, megaloblastic anemia and iron deficiency anemia
Henderson-Paterson inclusion	Molluscum contagiosum
Dilapidated brick wall effect	Familial benign chronic pemphigus
Cartwheel or checkerboard appearance	Multiple myeloma
Tobacco cells or cell in cells	Hereditary benign intra-epithelial dyskeratosis
Lava flowing around boulder'	Dentin dysplasia (shield type I)

Various drug effect on oral cavity

<i>Tissue</i>	<i>Drug effect</i>	<i>Drugs used</i>
Teeth	Discoloration Root anomalies	Tetracycline Chlorhexidine Phenytoin Cytotoxic
Gingiva	Swelling	Phenytoin Cyclosporine Nifedipine
Salivary gland	Dry mouth	Tricyclic antidepressant Phenothiazine Antihypertensive Lithium
Taste	Disturbed	Metronidazole Penicillamine
Facial movements	Dyskinesia	Phenothiazines Metoclopramide
Mucosa	Thrush Ulcers Lichenoid lesions Erythema multiforme	Broad spectrum antimicrobial Corticosteroids Cytotoxic drugs NSAID Barbiturates Sulphonamides

Summary of the main mandibular and maxillary fracture site and the common projection used for each side**Mandibular**

<i>Fracture site</i>	<i>Commonly used projection</i>
Angle	Dental panoramic tomography or oblique lateral Postero-anterior of jaws
Condylar neck	Dental panoramic tomography or oblique lateral Postero-anterior of jaws (for low neck fracture) Reverse Towne's (for high condylar neck)
Body	Dental panoramic tomography or oblique lateral Postero-anterior of jaws periapical of involved teeth, Lower 90° occlusal
Canine region	Dental panoramic tomography or oblique lateral Periapical of involved teeth, True lateral skull
Symphysis	Lower 45° occlusal, Lower 90° occlusal
Ramus	Dental panoramic tomography or oblique lateral Postero-anterior of jaws
Coronoid process	Dental panoramic tomography or oblique lateral 0° occipitomenal

Maxillary

<i>Fracture type/site</i>	<i>Commonly used radiograph</i>
Dento-alveolar	Periapical, Upper standard occlusal, Upper occlusal
Le Fort I	0° occipitomenal, 30° occipitomenal, True lateral skull (brow-up)
Le Fort II	0° occipitomenal, 30° occipitomenal, True lateral skull (brow-up)
Le Fort III	0° occipitomenal, 30° occipitomenal, True lateral skull (brow-up), Coronal section tomography, CT scan/3D reconstruction
Zygomatic complex	0° occipitomenal, 30° occipitomenal, Submentovertex
Nasoethmoidal	0° occipitomenal, 30° occipitomenal, True lateral skull (brow-up) Soft tissue lateral view of the nose, CT /3D reconstruction
Orbit	0° occipitomenal, True lateral skull (brow-up), Postero-anterior 25° CT/3-D reconstruction

Various lymph nodes in the head and neck region and their area of drainage

<i>Lymph nodes</i>	<i>Area of drainage</i>	<i>Discharge</i>
Occipital	Posterior scalp	Spinal accessory nodes
Retromandibular	Scalp, posterior ear, back of external auditory meatus	Internal jugular nodes
Superficial parotid	Lateral and frontal scalp, lateral ear, external auditory canal, eyelids	Superficial cervical nodes Internal jugular nodes
Deep parotid	Parotid gland, orbit, lateral eyelid, conjunctiva, superficial thyroid nodes	Internal jugular nodes
Buccal	Medial eyelids, skin and mucous membrane of the nose and cheeks	Mandibular nodes
Mandibular	Similar to buccal lymph nodes	Submandibular nodes
Submental	Tip of the tongue, anterior floor of mouth, anterior lower gingiva, middle lower lip, chin	Submandibular nodes
Submandibular	Salivary gland, upper and lower lips, cheeks, gingiva teeth, anterior palatine pillar, soft palate, anterior two-third of the tongue	Internal jugular nodes
Superficial cervical	Parotid nodes	Internal jugular nodes
Internal jugular	All the nodes mentioned above and pharynx, tonsil, tongue, palate, larynx	Subclavian vein, right side thoracic duct
Spinal accessory	Occipital nodes, retroauricular nodes, back of heads, nape and lateral aspect of the neck	Supraclavicular nodes
Supraclavicular	Spinal accessory nodes, posterior triangle submandibular nodes	Joins inferior internal jugular

Firm non-hemorrhagic soft tissue growth of oral cavity

- Fibromatosis
- Torus
- Irritation fibroma
- Peripheral fibroma
- Myxoma
- Neurofibroma
- Lipoma
- Granular cell myoblastoma
- Sialadenitis
- Tumor of salivary gland

Hemorrhagic or easily bleeding soft tissue growth of oral cavity

- Parulis
- Eosinophilic granuloma
- Epulis fissuratum
- Peripheral giant cell granuloma
- Pyogenic granuloma
- Pregnancy tumor
- Squamous cell carcinoma
- Lymphomas
- Leukemia
- AIDS

Compressible soft tissue growth of oral cavity

- Eruption cyst
- Mucocele
- Mucous cyst
- Ranula
- Gingival cyst
- Nasoalveolar cyst
- Epidermoid cyst
- Cavernous
- Capillary hemangioma
- Lymphangioma
- Cystic hygroma

Various odors related to systemic disease

- Sour—digestive dysfunction
- Fetid—ANUG
- Urinous—uremia, kidney dysfunction
- Acetone—diabetes mellitus
- Stench—gangrene, necrotic lesion
- Fruity—after use of ether for GA
- Mousy—chronic liver dysfunction
- Bloody—internal hemorrhage

Exophytic lesion						
Condition	Appearance	Fluctuance	Entitbility	Usual history	Age	Frequency
Black hairy tongue	Patch of hairy growth on dorsal surface of tongue	No	No	Feels like hair on tongue gagging sensation	Older than 40	Occasionally
Mucocele	Nodular swelling	Yes	No	Variation in size rupture and draining	<40	Occasional
Ranula	Nodular swelling on floor of mouth	Yes	No	Slowly enlarging smaller in early morning	—	Common in female
Cavernous hemangioma	Nodular swelling	Usually not	Yes	Present from birth	—	Occasional
Early hematoma	Same	Yes	No	Recent trauma and bleeding disorder	—	Occasional
Superficial hematoma	Same	Yes	No	Displace teeth slowly enlarging	—	Occasional
Lymphangioma	Nodular swelling pebbly appearance	Usually not	Partially	Present from birth and after trauma	—	Rare
Mucoepidermoid tumor	Mucocele like nodule	—	No	Slowly expanding mass	40	Rare
Multiple neuro-fibromatosis	Café au lait spot ion skin and oral mucosa	—	no	Multiple skin tumor from birth	—	Rare
Rendu-Osler-Weber syndrome	Purple papule on skin and mucus membrane which blanch on pressure	No	Yes	Bleeding from lesion and body orifice	12	Rare

Generalized discoloration

Condition	Distribution	History	Age	Accompanied condition	Special test
Cyanosis	Total skin surface and oral mucosa	Increased severity in excretion	1 month to 2 years	Malaise dyspnea	Color decreased when oxygenated blood increased
Chloasma gravidarum	Skin face over the nose and cheek	Increased brownish color of skin	Older than 13	Pregnancy	—
Addison disease	Skin exposed	Hypoglycemia weakness decreased resistance to stress	18 and older	Adrenocorticoids insufficiency	–ve ACTH test
Hemochromatosis	Skin exposed	Slowly increasing, increased iron uptake multiple blood transfusion older than 35	Liver disease, diabetes, adrenal insufficiency	Test of skin liver bone marrow +ve for iron	
Argyria	Same	Chronic self-administer of silver containing	18 and over	Equilibrium and hearing problems headache	Skin positive for silver

Papillary or cauliflower like soft tissue growth of oral cavity

- Verrucous leukoplakia
- Verruca vulgaris
- Condyloma acuminatum
- Papilloma
- Keratoacanthoma
- Inflammatory papillary hyperplasia
- Verrucous carcinoma

- Fine needle
- Punch
- Scrape
- Trephine

Less commonly used

- Bite
- Brush
- Cone
- Core
- Endoscopic
- Irrigation
- Pressure
- Shave
- Sponge

Types of biopsy**Commonly used**

- Aspiration
- Curettage
- Excisional
- Incisional

Differential diagnosis of bullous—erosive mucosal changes				
Feature	<i>Pemphigus</i>	<i>Bullous pemphigoid</i>	<i>Mucous membrane pemphigoid</i>	<i>Erythema multiforme</i>
Age of the patient	Under 50 years	Over 60 years	Over 40 years	Any
Oral mucosal involvement	100%	20%	100%	50-60%
Initial oral lesion	70%	3%	85%	45%
Lethal effect without therapy	90-100%	10-15%	Under 1%	Under 1%
Tzanck test	+ve	–ve	–ve	–ve
Immunofluorescence	Intercellular	Basal membrane	Basal membrane	–ve

Tabular presentation of tooth tissue loss

Term	Definition	Clinical Appearance
Attrition	Loss by wear of surface of tooth or restoration caused by tooth to tooth contact during mastication or parafunction	• Matching wear on occluding surfaces • Shiny facets on amalgam contact • Enamel and dentin wear at the same rate • Possible fracture of cusps or restorations
Abrasion	Loss by wear of dental tissue caused by abrasion by foreign substance (e.g. tooth-brush, dentifrice)	• Usually located at cervical areas of teeth • Lesions are more wide than deep • Premolars and cuspids are commonly affected
Erosion	Progressive loss of hard dental tissue by chemical processes not involving bacterial action	• Broad concavities within smooth surface enamel • Cupping of occlusal surfaces, (incisal grooving) with dentin exposure • Increased incisal translucency, wear on non-occluding surfaces and raised amalgam restorations • Clean, non-tarnished appearance of amalgams • Loss of surface characteristics of enamel in young children • Preservation of enamel "cuff" in gingival crevice is common • Hypersensitivity and pulp exposure in deciduous teeth
Abfraction	Loss of tooth surface at the cervical areas of teeth caused by tensile and compressive forces during tooth flexure	• Affects buccal/labial cervical areas of teeth • Deep, narrow V-shaped notch • Commonly affects single tooth with excursive interferences or eccentric occlusal loads

Causes and mechanism of vitamin D deficiency

Predisposing factors	Mechanism
Dietary lack of meat and dairy product	Low levels of vitamin D in the diet
Lack of adequate exposure to ultraviolet light	Failure of vitamin D precursor synthesis in the skin
Gastric intestinal disease or chronic liver disease	Malabsorption of vitamin D and calcium
Aluminum toxicity and biphosphonates	Direct inhibition of bone mineralization
Administration of anticonvulsant drug like phenobarbitone	These drugs enhance liver enzyme activity which result in increase breakdown of vitamin D to biological inert product
Chronic renal failure	Reduced conversion of 25(OH)D ₃ to 1,25 (OH) ₂ D ₃
Hypophosphatemia rickets (X linked dominant)	Inherited defect in renal tubular phosphate reabsorption leading to hypophosphatemia
Hypophosphatasia (autosomal recessive)	Defect mutation in bone alkaline phosphatase which cause inhibition of bone mineralization at the calcification front

Radiological features of periapical cemental dysplasia

Stage	Appearance	Notes
Stage 1 Radiolucent (fibrous)	Identical to that seen in routine periapical rarefying osteitis	Vitality test of involved teeth is mandatory during this stage
Stage 2 Mixed stage	Minute radiopacities appear in the radiolucent periapical lesion	As this stage progress, the small radiopacities may coalesce The overall radiolucency usually does not enlarge further
Stage 3 Radiopaque (calcifying stage)	Substantial or complete opacification A radiolucent capsule (which may be very thin) always surrounds the opacity and separates it from the adjacent normal bone	The radiolucent capsule is of considerable diagnostic significance

Comparison of different clinical forms of histiocytosis-X				
Current designation	Former designation	Age of occurrence	Lesion distribution	Clinical course
Acute disseminated histiocytosis	Letterer-Siwe disease	Infancy and early childhood	Multiple lesion affecting bone, skin and other organs	Rapidly progressive often fatal; considered by many to be malignant neoplastic disease
Chronic disseminated idiopathic histiocytosis	Hand-Schuler-Christian disease	Children	Multiple bone lesion and extra-bony manifestation such as exophthalmos, diabetes insipidus, lymphadenopathy, splenomegaly, hepatomegaly, dermatitis	Gradual progression, lesion usually controlled by surgical removal and/ or low dose radiotherapy
Chronic localized idiopathic histiocytosis	Eosinophilic granuloma	Older children, adolescents and young adults	Isolate or multiple lesion limited to bone	Manifestation considered as either idiopathic inflammatory or benign neoplastic disease; usually controlled by surgical removal, although progression into the chronic disseminated form can occur

Different types of oral drug-reaction patterns and associated drugs	
Reaction	Drugs responsible
Xerostomia	Antidepressants and antipsychotics, antihypertensives, antihistamines, anticholinergics, and decongestants
Swelling	Penicillins, aspirin, and ACE inhibitors
Nonspecific ulceration	Antineoplastics, including methotrexate, 5-fluorouracil, doxorubicin, and melphalan; barbiturates; dapsone; phenazone derivatives; phenolphthalein; salicylates; sulfonamides; tetracycline; and (direct contact with) compounds containing aspirin, hydrogen peroxide, or phenol
Lichen planus-like	Allopurinol, amiphenazole, amphotericin B, antimalarials (chloroquine, hydroxychloroquine, quinacrine, quinidine), arsenicals, beta-blockers, bismuth, captopril, carbamazepine, chlorothiazide, chlorpropamide, cimetidine, cinnarizine, cyanamide, dapsone, fenclofenac, flunarizine, furosemide, gold salts, isoniazid, ketoconazole, levopromazine, levamisole, lithium, lorazepam, mercury, methylidopa, methopromazine, oxprenolol, palladium, para-aminosalicylic acid, penicillamine, phenothiazines, phenylbutazone, practolol, propranolol, pyrimethamine, pyritinol, spironolactone, sulfonylureas, streptomycin, tetracycline, tolbutamide, and triprolidine
Erythema multiforme-like	Antibiotics (antimalarials, penicillins, sulfonamides), barbiturates, and salicylates
Pemphigoid-like	Antirheumatics (penicillamine, ibuprofen, phenacetin), cardiovascular drugs (furosemide, captopril, clonidine), antibiotics (penicillins, sulfonamides), antimicrobials, thiol-containing drugs, and sulfonamide derivatives
Pemphigus-like	Alpha-mercaptopyrionylglycine, ampicillin, captopril, cephalixin, ethambutol, glibenclamide, gold, heroin, ibuprofen, penicillamine, phenobarbital, phenylbutazone, piroxicam, practolol, propranolol, pyritinol chlorohydrate, rifampin, and theobromine
Lupus-like	Carbamazepine, chlorpromazine, ethosuximide, gold, griseofulvin, hydantoins, hydralazine, isoniazid, lithium, methylidopa, penicillamine, primidone, procainamide, quinidine, reserpine, streptomycin, thiouracils, and trimethadione
Nonspecific vesiculoulcerative mucositis	Nonsteroidal anti-inflammatory drugs (NSAIDs) (i.e. indomethacin, gold salts, naproxen), meprobamate, methylidopa, penicillamine, phenylbutazone, propranolol, spironolactone, thiazides, and tolbutamide
Pigmentation	Amiodarone, antimalarials (chloroquine, hydrochloroquine, hydroxychloroquine, quinacrine, quinidine), busulfan, clofazimine, cyclophosphamide, estrogen, ketoconazole, minocycline, phenolphthalein, tranquilizers (chlorpromazine), and zidovudine
Gingival enlargement	Calcium channel blockers (amlodipine, bepridil, diltiazem, felodipine, isradipine, nicardipine, nifedipine, nimodipine, nisoldipine, nitrendipine, oxidipine, verapamil), other dihydropyridines (bleomycin), cyclosporine, phenytoin, and sodium valproate

Various drug reaction in oral cavity	
<i>Reaction</i>	<i>Drugs</i>
Xerostomia, or dry mouth	Antidepressants, antipsychotics, antihypertensives, antihistamines, and anticholinergics
Swelling	ACE inhibitors, penicillin and penicillin derivatives, cephalosporins, barbiturates, and aspirin and other NSAIDs
Osteonecrosis of the jaws	Bisphosphonate (bp) therapy
Gingival enlargement	Phenytoin, calcium channel blockers (members of the dihydropyridine class of medications), cyclosporine, and the antiepileptic drug sodium valproate within the calcium channel blocker family, nifedipine, diltiazem, verapamil, and amlodipine are among the most commonly reported causative agents
Pigmentation	Antimalarial agents, particularly chloroquine, hydroxychloroquine, quinacrine, and quinidine tranquilizers, especially chlorpromazine, zidovudine, clofazimine, and ketoconazole, Minocycline
Nonspecific vesiculoulcerative mucositis	Thiazide derivatives, naproxen, gold salts, and penicillamine
Lupus-like reactions	Procainamide and hydralazine
Pemphigus-like reactions	Thiol-containing drugs are the most common cause of pemphigus-like reactions
Pemphigoid-like reactions	Thiol-containing drugs and sulfonamide derivatives are among the most commonly involved medications, as are the therapeutic classes of Nsaids, cardiovascular agents, antimicrobials, and antirheumatics
Lichen-planus-like or lichenoid reactions	Antimalarial medications NSAIDs and ACE inhibitors
Erythema multiforme like reactions	Sulfonamides, sulfonylureas, and barbiturates
Nonspecific ulceration	Aspirin, hydrogen peroxide, potassium tablets, and phenol-containing compounds barbiturates, beta-blockers, dapsone, NSAIDs, phenazone derivatives

2

Syndromes of Oral Cavity

Adaptation Syndrome

- It is disease of hormone which respond to stimulation.
- Patient may suffer from hypertension, periarteritis nodosa and other hormone related disorders.

Adrenogenital Syndrome

- It occurs due to hyperplasia or tumors of renal cortex.
- Pseudohermaphroditism, sexual precocity and virilism in women or feminization in men.
- *Oral features*—premature eruption of teeth if the disease begins in early life.

Aglossia-Adactylia Syndrome

- Complete absence of tongue.
- Focal enlargement of salivary gland usually in hard palate area.

Albright's Syndrome

- It is also called as 'McCune-Albright syndrome'.
- Severe fibrous dysplasia involving nearly all bones of the skeleton.
- Pigmented lesions of the skin (*cafe' au lait* spots).
- *Endocrine disturbances*—hyper-function of one or more endocrine glands.

Aldrich's Syndrome

- It is also called as 'Wiskott-Aldrich' syndrome.
- It is characterized by thrombocytopenic purpura, eczema, and increased susceptibility to infection.
- *Oral features*—spontaneous bleeding from gingiva and palatal petechiae can be seen.

Amelo-onycho-hypohidrotic Syndrome

- Defective nails and hypofunction of the sweat glands. There is also seborrheic dermatitis.
- *Oral features*—severe hypoplastic hypocalcified enamel.

Anderson Syndrome

- It is also called as '*familial osteodysplasia*'.
- Craniofacial and skeletal anomalies.

- Presence of diastolic hypertension and hyperuricemia.
- Oral features include maxillary hypoplasia, reduced ramus, mandibular prognathism and malocclusion.

Angio-osteo-hypertrophy Syndrome

- Port-wine stain on the face, varices.
- Hypertrophy of bone including jaw bone.
- *Oral features*—facial asymmetry, malocclusion and altered eruption pattern of teeth.

Aortic Arch Syndrome

- It is also called as '*pulseless disease*'.
- It is caused by narrowing or obstruction of the major branches of the arch of the aorta.
- *General features*—dizziness, headache, visual disturbance and anginal pain.
- *Oral features*—there is tropic ulceration and pain while chewing. It is result from deficient blood supply to the muscle of mastication.

Apert's Syndrome

- It is also called as '*acrocephalosyndactyly*'.
- *Skeletal deformities*—there is syndactyly (fusion of finger) of second, third and fourth digit of hand and acrobrachycephaly (tower skull). In some cases kleeblattschadel deformity (cloverleaf skull). The skull is ovoid, brachycephalic and often presents a horizontal supraorbital groove.
- *Facial deformities*—the middle-third of face is undeveloped.
- *Oral features*
 - High palatal vault and V-shaped maxillary alveolar ridge.
 - Trapezoid shaped appearance of lip when lip are relaxed.
 - There is posterior palatal cleft and bifid uvula.
 - Retarded eruption and dental malocclusion.
- Class II malocclusion.

Ascher's Syndrome

- Patient is having a double lip.
- Blepharochalasis, i.e. drooping of the tissue between the eyebrow and the edge of upper eyelid.
- Non-toxic thyroid enlargement.

Auriculotemporal (Frey's) Syndrome

- It is caused by damage to auriculotemporal nerve.
- Flushing and sweating of the involved side of face, chiefly in temporal area, during eating.
- Gustatory sweating when eating spicy food.

B-K Mole Syndrome

- It is autosomal dominant condition.
- It is characterized by large pigmented nevi.
- There is high-risk of development of melanoma.

Baby Bottle Syndrome

- It is also called as 'nursing bottle caries' or 'bottle mouth syndrome'.
- It occurs due to habitual use of bottle usually as an aid for sleeping in night.
- There is wide spread carious destruction of deciduous teeth, most commonly the four maxillary incisors followed by the first molars and then the cuspids if habit prolonged.

Behçet's Syndrome

- *Recurrent oral ulceration*—it has similar appearance as aphthous ulcer.
- *Recurring genital ulceration*—ulcer of scrotum and penis in males and ulcers of labia in females.
- *Skin lesions*—they are manifested as large pustular lesions.
- *Ocular lesions*—it consist of uveitis, retinal vasculitis, optic atrophy, recurrent conjunctivitis and keratitis.

Beckwith's Hypoglycemic Syndrome

- *Macroglossia*—enlargement of the tongue.
- *Other features*—it includes neonatal hypoglycemia, mild microcephaly, umbilical hernia, fetal visceromegaly and postnatal somatic gigantism.

Bernard-Soulier Syndrome

- It is transmitted as autosomal dominant trait with variable penetration.
- The membrane receptors on platelets are absent and it accounts for bleeding problems.
- The bleeding time is prolonged.

Blepharochelodontic Syndrome

- It is transmitted as autosomal dominant inheritance.
- *Eye anomalies*—it includes lagophthalmus, ectropion of lower eyelid.
- *Lip*—there is bilateral cleft lip and palate.
- *Teeth*—oligodontia, microdontia including tiny molars.

Bloch-Sulzberger Syndrome

- Erythematous and vesiculobullous lesions on the trunk and extremities.
- These are replaced by white keratotic, lichenoid, papillary or verrucous lesions.
- Brownish gray macules in a streaked, patchy distribution over the trunk and extremities.

- *Oral features*—delayed tooth eruption, peg or cone shaped teeth, congenitally missing teeth, malformed teeth and additional cusps.

Blepharonasofacial Syndrome

- It is characterized by mental retardation, joint disorders and craniofacial anomalies.
- *Facial features*—affected individuals show microcephaly, an anti-mongoloid slant of palpebral fissures.
- *Oral features*—there is also hypoplastic maxilla, protruding lip and malocclusion resulting from midface hypoplasia.

Book's Syndrome

- Premature whitening of hair.
- Hyperhydrosis of palms and soles.
- *Oral features*—absence of the premolars and third molars.

Bowen Syndrome

- It is also called as 'cerebrohepatorenal syndrome'.
- Craniofacial anomalies, hypotonia, hepatomegaly and renal cortical cysts.
- *Oral features*—it includes micrognathia, protruding tongue and high arched palate.
- There is increase serum iron level and decrease in serum immunoglobulin levels.

Branchial Arch Syndrome

- It is heterogeneous group of malformation of the head and neck characterized by anatomic alteration in the structure which is derived from the branchial arches.

Burning Mouth Syndrome

- Pain and burning sensation in the mouth.
- Altered taste sensation and xerostomia.
- No clinically detectable lesions in the oral cavity.

Caffey-Silverman Syndrome

- It is also called as 'infantile cortical hyperostosis'.
- Development of tender deeply placed soft tissue swellings and cortical thickening or hyperostosis involving various bones of the skeleton.
- There is also pain, fever and irritability in infants.
- Increased serum levels of alkaline phosphatase and increased ESR.

Carotid Artery Syndrome

- Deviated styloid process or ossified ligament causing impingement on the internal or external carotid artery exerting pressure.
- There is atypical facial pain.

Candidosis Endocrinopathy Syndrome

- *Oral features*—chronic oral candidiasis and enamel hypoplasia.
- *Endocrine disorders*—it includes hypoparathyroidism, hypothyroidism, and hypo-adrenocorticism.
- *Metabolic disorders*—diabetes mellitus.

Carpenters Syndrome

- *Cloverleaf skull*—often associated with this defect is marked cranial asymmetry described as a “cloverleaf” skull anomaly caused by the craniosynostosis.
- *Eyes*—there is mild down sloping of the eyes, epicanthal folds, as well as malformations of the eyes themselves.
- *Ears*—the ears of these patients are low set, their necks are short.
- *Mandible*—mandible may be somewhat small.
- *High arched palate*—commonly seen in these patients is a highly arched and somewhat narrow palate.
- *Mental deficiency*—the mental deficiency seen with these patients varies from mild to severe, although multiple cases have been reported with normal intelligence.
- *Other*—tower shaped skull, presence of additional or fused fingers and toes, reduced height and obesity.

Cerebrocostomandibular Syndrome

- Thoracic deformity with barking cough and mental retardation.
- *Oral features*—it includes mandibular micrognathism, palatal defect and absence of uvula or even soft palate.

Carcinoid Syndrome

- Intermittent attack of flushing of skin of face.
- Watery diarrhea, abdominal pain, attacks of dyspnea.
- Right side heart failure due to involvement of tricuspid and pulmonary valve and endocardium.

Chediak-Higashi Syndrome

- It is transmitted as autosomal dominant trait.
- *General*—it includes oculocutaneous albinism, photophobia, nystagmus and recurrent infection of respiratory tract and skin.
- *Oral*—ulceration of the oral mucosa, severe gingivitis and glossitis.

Costen's Syndrome

- There is impairment of hearing, either continuously or intermittent.
- A stuffy sensation in the ears especially at mealtime.
- Tinnitus, otalgia, dizziness and headache about the vertex, occiput and behind the ears.
- Burning sensations in the throat, tongue and side of nose.

Cowden's Syndrome

- It is autosomal dominant disease characterized by facial trichilemmomas associated with gastrointestinal tract, central nervous system, thyroid and musculoskeletal abnormalities.
- Papillomatous lesion as well as ‘pebbly’ lesions of lip, gingivae, palate and pharynx occurs.
- Fibromas at various sites in the oral cavity.
- Lichenoid and papillomatous lesions of perioral, perinasal and periorbital areas of ear and neck.
- Hematoma of skin, gastrointestinal tract, breast and thyroid.

Cracked Tooth Syndrome

- Development of crack in a restored or unrestored tooth due to excessive occlusal force.
- Sharp pain on biting.

Cross Syndrome

- *Oral features*—it include gingival enlargement.
- *Other features*—it includes hypopigmentation, oligophrenia, microphthalmos and athetosis.

CREST Syndrome**Associated with scleroderma**

- C—Calcinosis cutis
- R—Raynaud's phenomenon
- E—Esophageal dysfunction
- S—Sclerodactyly
- T—Telangiectasia

Crouzon Syndrome

- It is also called as ‘craniofacial dysostosis’.
- *Cranial deformities*—protuberant frontal region with an anteroposterior ridge overhanging the frontal eminence and often passing to the roof of nose (triangular frontal defect). The cranium is brachycephalic.
- *Facial malformations*—there is hypoplasia of maxilla with mandibular prognathism.
- *Parrot beak*—the upper lip is short and nose resembles ‘Parrot's beak’.
- Oral feature includes high arched palate, V-shaped dental arch, peg shaped teeth and partial anodontia.
- *Eye changes*—hypertelorism, exophthalmos with divergent strabismus, optic neuritis and choked disc resulting frequently in blindness.
- *Others*—spina bifida occulta.

Cushing's Syndrome

- It is characterized by adiposity about the upper portion of the body, mooning of the face and tendency to become round shouldered.
- *Buffalo hump*—it is seen at the base of the neck.
- There is dusky plethoric appearance with formation of purple striae.
- There is also muscular weakness, vascular hypertension, glycosuria and albuminuria.
- *Children*—there may be osteoporosis and premature cessation of epiphyseal growth.

Curry-Hall Syndrome

- Short limbs, polydactyly and nail dysplasia.
- *Oral features*—deciduous teeth are small and conical. Incisors are retained as permanent successors may be congenitally missing.

Dejerine-Roussy Syndrome

- Tumors of the pons or occlusion of the posterior cerebral artery with sensory or motor abnormality on the contralateral side.
- *Oral features*—orofacial pain and dysgeusia are also present.

Down's Syndrome

It is also called as 'trisomy 21 syndrome' or 'mongolism'.

Systemic

- **Cardiovascular**—there is ventricular septal defect, ALV communication, patent ductus arteriosus and mitral valve prolapse.
- **Hematological features**—it includes impaired immuno-deficiency, short lived neutrophils, risk of lymphopenia, eosinophilia, increased risk of leukemia and increased risk of hepatitis carrier state.
- **Musculoskeletal**—it includes atlantoaxial instability; midface is underdeveloped with relative prognathism, narrow and partially obstructed nasal passage and open mouth with tongue thrusting habit.
- **Nervous**—motor function is delayed, dementia analogues and phonation distorted.
- **Behavior**—gentleness, anxiety and stubbornness.

Oral

- **Palate**—V-shaped high vault and soft palate insufficiency.
- **Oral opening**—angle of mouth is pulled down due to hypotonic musculature, lower lip is everted, mouth breathing with drooling of saliva, chapped lower lip and angular cheilitis.
- **Tongue**—scalloped or fissured tongue, protrusion with tongue thrusting, macroglossia and desiccated tongue.
- **Teeth**—microdontia, hypodontia, partial anodontia, supernumerary teeth, spacing, taurodontism, crown variation, agenesis, hypoplasia and hypocalcification and delayed eruption of teeth.
- **Occlusion**—malalignment, frequent malocclusion, frequent TMJ dysfunction and bruxism.

Eagle's Syndrome

- It is characterized by elongation of styloid process or ossification of stylohyoid ligament.
- **Features**—it includes dysphagia, sore throat, otalgia, glossodynia, headache and vague orofacial pain.

Edward's Syndrome

- It is also called as 'trisomy 18 syndrome'.
- The affected individuals are mentally retard and show hypertonicity.
- **Facial features**—there are small eyes and prominent occiput.
- The index finger overlaps the 3rd finger and 5th finger overlaps the 4th finger.
- **Oral features**—it includes micrognathia, high arched palate, cleft palate and bifid uvula.

Ehlers-Danlos Syndrome

- It is also called as 'rubber man' and is autosomal dominant.
- **General**—there is hyperelasticity of skin, hyperextensibility of joint and fragility of skin and blood vessels.
- **Oral features**—it includes enamel hypoplasia and periodontal destruction is severe. There is also hypermobility of TMJ resulting in repeated dislocation.

Elashy Water's Syndrome

- It is also called as 'brachioskeleto-genital syndrome'.
- There is mental retardation, brachycephaly face, divergent strabismus and ocular hypertelorism.
- **Oral features**—it includes cleft palate and multiple jaw cysts.

Ellis-van Creveld Syndrome

- It is also called as 'chondroectodermal dysplasia'. It is inherited as autosomal recessive trait.
- **Skeletal deformities**—the affected person is of short stature due to chondrodysplasia. Polydactyly is also reported.
- **Cardiac anomalies**—congenital heart disease may cause neonatal death.
- **Hair and nails defect**—nails are dystrophic. Eyebrows and pubic hair are often deficient.
- **Oral features**
 - Fusion of middle portion of the upper lip to maxillary gingival margin eliminating the normal upper labial sulcus.
 - Deciduous teeth show hypoplasia and permanent tooth eruption is delayed.
 - Accessory cusp is common.

Epidermis Nevus Syndrome

- Cutaneous nevi extending upto oral mucosa and gingiva.
- Mental deficiency, skeletal abnormality.
- There is also presence of hypoplastic teeth.

Fanconi's Syndrome

- Congenital or familial aplastic anemia.
- Bone abnormalities, microcephaly and generalized olive-brown pigmentation of the skin.

Favre-Racouchot Syndrome

- It occurs due to ultraviolet light and excessive smoking habit.
- There is extensive sun damage of the facial skin.
- There are also numerous open, dilated and cystic comedones.
- Dermis show solar elastosis characterized by dilated pilosebaceous opening with distended, horn filled hair follicle.

First Arch Syndrome

- **Oral features**—it includes cleft lip and palate and mandibulofacial dysostosis.
- Hypertelorism and deformities of ear.

Floppy Infant Syndrome

- Generalized weakness due to hypotonia.
- Inability to sit, stand and walk.
- Hypotonia may involve tongue and facial muscles.

Fragile X Syndrome

- X-linked mental retardation, macro-orchidism and large ears.
- Long narrow face and cleft palate.
- Mitral valve prolapsed.

Focal Dermal Hypoplasia Syndrome

- Focal absence of dermis associated with herniation of subcutaneous fat into the defects.
- Skin atrophy, streaky pigmentation, telangiectasia.
- Multiple papilloma of skin or mucosa, syndactyly, polydactyly and adactyly.
- Asymmetrical face with pointed chin and notched nasal alae, papillomas present on lips, buccal mucosa and gingiva.
- Microdonia with defect in size of teeth.
- Enamel hypoplasia, cleft lip/cleft palate.

Franceschetti's Syndrome

- Antimongoloid palpebral fissure with colobomas of the outer portion of the lower eyelids and deficiency of eyelashes.
- Hypoplasia of facial bone especially malar bone mandible.
- Malformation of external ear and middle ear.
- High palate and malocclusion is also present.
- Blind fistulae between angle of the ears and angle of mouth.
- Typical hair growth in form of tongue shape extending towards the cheek.
- Bird like or fish like appearance.

Gardner's Syndrome

- *Systemic features*—multiple polyposis of large intestine and polyp of colon and rectum.
- *Tumors*—osteomas of bone including long bones, skull and jaws. There may be occasional occurrence of desmoid tumors. Other tumors which can occur are lipoma, leiomyoma and adenocarcinoma of colon.
- *Cysts*—multiple epidermoid or sebaceous cyst of skin particularly of scalp and back.
- *Oral features*—hypercementosis, multiple unerupted supernumerary teeth and compound odontoma.

Gilles de la Tourette's Syndrome

- Spontaneous erratic behavior of the patients.
- Incoherent facial expressions and verbalization.
- Tendency for self-mutilation of oral tissue by use of teeth and finger nails.

Goltz-Gorlin Syndrome

- It is also called as 'focal dermal hypoplasia syndrome'. It is transmitted as an autosomal dominant trait.
- *General features*
 - Focal absence of dermis associated with herniation of subcutaneous fat into the defects.
 - There is also skin atrophy, streaky pigmentation and telangiectasia.
 - Multiple papillomas of skin or mucosa.
 - Syndactyly, polydactyly, and adactyly.
- *Oral features*—papillomas of lip, microdonia, cleft lip and cleft palate.

Gorlin-Chaudhry-Moss Syndrome

- It is characterized by craniofacial dysostosis, patent ductus arteriosus, hypertrichosis and hypoplasia of labia majora.
- Oral feature of this syndrome is hypodontia.

Goldenhar's Syndrome

- Unilateral microstomia, mental retardation, hypoplastic zygomatic arch.
- *Facial features*—there is downward slanting of palpebral fissures, malformed pinna and iris coloboma.
- *Oral features*—there is high arched palate, palatal and uvular cleft with malocclusion.

Gorham's Syndrome

It also called as 'massive osteolysis' or 'Phantom bone'.

- Osteolysis of single or multiple bones followed by replacement with fibrous tissue.
- Pain in the bone and pathological fractures.
- *Oral features*—there may be destruction of mandible or maxilla. Pain and facial asymmetry can be seen.

Graham Little Syndrome

- It combines small confluent patches of progressive scarring alopecia resembling that of lichen planus follicularis.
- There is also follicular keratosis, and non-cicatricial alopecia of the axilla and pubes.
- Patches on the scalp are atrophic and cicatricial in the center but erythematous and squamous around the edges, where follicular keratosis occurs when the disease is actively progressing.

Grinspan Syndrome

- It is triad of lichen planus, diabetes mellitus and vascular hypertension.

Gradenigo's Syndrome

- Suppurative otitis
- Trigeminal nerve pain
- Abducens nerve palsy

Hajadu Cheney Syndrome

It is rare autosomal dominant disorder characterized by short stature, disintegration of terminal phalanges of fingers and toes.

- *Facial features*—long nose, low frontal hairline and flared ears.
- *Skeletal feature*—the individual is usually victim of multiple fractures of bones. Sutures are usually open and frontal sinuses are absent.
- *Oral features*—there is premature loss of teeth and abnormal shape of skull.

Hanhart's Syndrome

- *Digital deformities*—it includes oligodactyly, syndactyly and hypoplastic digits.
- *Oral features*—there is micrognathia, microglossia and hypodontia.

Hallermann-Streiff Syndrome

- *Skeletal and genital*—it includes syndactyly and hypogenitalism.
- *Facial features*—it includes microphthalmia, long thin tapering nose, strabismus, and double cutaneous chin with central furrow, hypotrichosis of the scalp and eyebrows and prominent scalp veins.

- *Oral features*—it includes hypodontia, microstomia, malocclusion, malformed teeth and retained deciduous teeth.

Heerfordt's Syndrome

- Firm painless, bilateral enlargement of parotid gland.
- Inflammation of uveal tract of the eye.
- Facial palsy.

Hetch-Beals-Wilson Syndrome

- Limited mandibular opening.
- Shortened leg, hamstring muscles and club foot.

Horner's Syndrome

- Miosis or contraction of pupil of the eye due to paresis of dilators of pupil.
- Ptosis or drooping of eyelid due to paresis of smooth muscle elevators of upper lid.
- Anhidrosis and vasodilatation over the face due to interruption of sudomotor and vasomotor control.

Horton's Syndrome

- It is also called as '*sphenopalatine neuralgia*'.
- Unilateral paroxysms of intense pain in the eye, maxilla, ear, mastoid region, base of nose and beneath the zygoma.
- Absence of trigger zones and occurrence of pain everyday exactly at the same time- for this reasons it is called as '*alarm clock headache*'.

Hurler's Syndrome

- It is disturbance in mucopolysaccharide metabolism.
- *Facial features*—prominent forehead, broad saddle nose, wide nostrils, hypertelorism and puffy eye lids. There is also corneal clouding present.
- There is also nasal congestion with noisy breathing.
- '*Claw hand*' resulting from flexion contractures.
- Hepatosplenomegaly results in protuberant abdomen.
- Death usually occurs before the age of 10 years.
- *Oral features*—thick lips, large tongue, open mouth.

Hunter's Syndrome

- Similar feature like Hurler's syndrome but they are mild.
- Corneal clouding is absent.
- Death usually occurs before the age of 15 years.

Hutchinson-Gilford Syndrome

- It is also called as '*progeria*'. It is transmitted as autosomal dominant trait. There is manifestation of old age features, pro- before, geria- old.
- Alopecia, pigmented areas of trunk, atrophic skin, prominent vein and loss of subcutaneous fat.
- The individual has high pitched, squeaky voice and beak-like nose.
- *Oral features*—loss of all teeth in very young age, hypoplastic mandible, and delayed eruption of teeth can also occur.

Hypoglossia-hypodactylia Syndrome

- Total or partial absence of the tongue.
- Micrognathia, high arched or cleft palate, glossopalatine ankylosis, defects in the lower lip and hypertrophy of sublingual glands.
- Hypodactylia and hypomelia.

Jaw-Winking Syndrome

- It is also called as '*marcus gun phenomenon*'.
- Rapid elevation of the ptotic eyelid occurring on movement of the mandible on the contralateral side.
- Congenital unilateral ptosis.

Jadassohn-Lewandowsky Syndrome

- Bilateral oral white lesions involving the tongue and buccal mucosa.
- Laminated thickening of fingers and toe nails.

Jaw Cyst-Basal Cell Nevus-Bifid Rib Syndrome

- It is also called as '*Gorlin-Goltz syndrome*'.
- *Cutaneous anomalies*—basal cell carcinoma, dermal cysts and tumors, palmar pitting, palmar and plantar keratosis and dermal calcinosis.
- *Dental abnormalities*—odontogenic keratocysts and mild mandibular prognathism.
- *Osseous abnormalities*—bifid rib, vertebral anomalies and brachy-metacarpalism.
- *Ophthalmologic anomalies*—hyper-telorism with wide nasal bridge, dystopia canthorum, congenital blindness and internal strabismus.
- *Neurological anomalies*—mental retardation, agenesis of corpus callosum, ductal calcification, congenital hydrocephalus and medulloblastomas.
- *Sexual anomalies*—hypogonadism, ovarian tumors.

Jugular Foramen Syndrome

- Dysphagia, hoarseness of voice, glossopharyngeal neuralgia like pain.
- Palatal weakness and vocal cord paralysis.

Kartagener's Syndrome

- Bronchiectasis
- Situs in versus
- Sinusitis

KBG Syndrome

- The affected individual show short stature and mental retardation.
- *Facial features*—flat bridge of nose, round face, long philtrum and abnormal auricle, with or without hearing defect.
- *Oral features*—include macrodontia, oligodontia, hypoplasia of enamel and micrognathia.

Klinefelter's Syndrome

- It occurs in males whose sex chromosome constitution includes one or more extra chromosomes.
- The patient develops infertility and gynecomastia.

- There may be fatigue, osteoporosis, mitral valve prolapse and varicose veins.
- The patients develop taurodontism.

Klippel-Trenaunay-Weber Syndrome

- *Hemangioma*—hemangioma of facial region.
- *Ocular lesion*—bluish, thin sclera, glaucoma.
- *Neurological symptoms*—epilepsy, mental retardation.
- *Oral finding*—gingival enlargement can be present.

Larsen's Syndrome

- It is an autosomal dominant disorder.
- There is prominent forehead with frontal bossing, flattened midface, depressed nasal bridge and hypertelorism.
- There is bilateral anterior dislocation of tibia or femur with displaced patella.
- *Oral features*—it includes cleft palate and malocclusion.

Laugier-Hunziker Syndrome

- Acquired pigmented macules in the lips, oral cavity and fingers.

Lowe's Syndrome

- Generalized aminoaciduria, mental retardation, hypotonia, congenital cataracts.
- Abnormal skull shaped.

Maffucci's Syndrome

- Multiple chondroma of the jaw bone.
- Multiple hemangiomas of skin and oral mucosa. Phleboliths can also occur.
- In oral mucosa, it occurs on the tongue.

Marfan's Syndrome

- *Skeletal*—excessive length of tubular bone resulting in disproportionate long thin extremities.
- *Craniofacial*—skull and face are long and narrow. Ears are large, eyes appear sunken and frontal bossing is seen.
- *Ocular*—ocular lens subluxation with a defect in the suspensory ligament.
- *Cardiovascular*—aortic aneurysm and regurgitation.
- *Oral*—temporomandibular joint dysarthrosis, multiple odontogenic cysts of maxilla and mandible, high arched palate and bifid uvula.

Magic Syndrome

- Mouth and genital ulcers with inflamed cartilage.

Marin Amat's Syndrome

- It is also called as '*inverted marcus gun phenomenon*'.
- Eye closes automatically when patient opens his mouth forcefully and fully, tears may also follow.

Melnick-Needles Syndrome

- It is autosomal dominant condition characterized by generalized bony dysplasia and abnormal facies.

- There is marked exophthalmos, full cheeks and large ears.
- *Oral features*—micrognathia, transversely long mouth and malocclusion.
- *Skeletal*—delayed closure of anterior fontanelle, defect in clavicles, ribs and vertebrae.

Median Cleft Face Syndrome

- It is also called as '*frontonasal dysplasia*'.
- There is nasal clefts and notches, preauricular tags and ocular hypertelorism.
- There is also median cleft of premaxilla and palate and malocclusion.

Melkersson-Rosenthal Syndrome

- It is triad of cheilitis granulomatosa, facial paralysis and scrotal tongue.

Miescher's Syndrome

- It is also called as '*cheilitis granulomatosa*'.
- Diffuse swelling of lip especially lower lip.
- Scarring, fissuring, vesicle or pustule formation on the vermillion border.
- It is associated with Melkersson-Rosenthal syndrome.

Middle Fossa Syndrome

- It occurs due to tumor in the region of gasserian ganglion.
- There is hyperesthesia, paraesthesia and paralysis of ocular muscles.
- Deviated mandibular opening and unilateral soft palate paralysis.

Mobius Syndrome

- It is also called as '*congenital facial diplegia*'.
- In infancy, failure to close the eyes during sleep.
- Partial/complete facial paralysis results in no change in facial expressions while crying or laughing.
- Drooling of saliva and difficulty in mastication.
- There is also external ophthalmoplegia, deformity of external ear, deafness, defect of pectoral muscle, paresis of tongue, soft palate or jaw muscles, clubfoot, mental defects and epilepsy.

Mohr's Syndrome

- It is autosomal recessive disorder characterized by several oral, facial and digital defects. The affected individual is moderately short.
- *Digital deformities*—brachydactyly, syndactyly or polydactyly.
- *Facial deformities*—midline cleft lip and bifid tip of nose.
- *Oral features*—there is high arched palate, lobate tongue, hypoplastic body of the mandible and hypodontia.

Morquio's Syndrome

- Severe enamel hypoplasia with grey and pitted enamel.
- Severe bone changes, corneal clouding and aortic regurgitation.

Multiple Endocrine Neoplasia Syndromes**MEN 1**

- Tumor or hyperplasia of pituitary; parathyroid gland, adrenal cortex and pancreatic islets.
- There is also peptic ulcer and gastric hypersecretion.

MEN 2 (Sipple's syndrome)

- Parathyroid adenoma or hyperplasia but no tumor of pancreas.
- Pheochromocytomas of adrenal medulla and medullary carcinoma of thyroid gland occurs.

MEN 3**Systemic**

- Mucocutaneous neuromas, pheo-chromocytomas of adrenal medulla and medullary carcinoma of thyroid.
- *Marfanoid habitus*—it refers to a slender body build, with long, thin extremities and increased laxity of joints. The face appears long and thin.

Oral

- Neuromas of lip, tongue and buccal mucosa.
- Thick and bumpy lips and infrequent prognathism.

Murray Puretic-Dresher Syndrome

- Gingival fibromatosis.
- Multiple hyaline fibromas of head, trunk and extremities.
- Suppurative skin lesions and flexion contractures.

Mucocutaneous Lymph Node Syndrome

- Bilateral congestion of ocular conjunctive and edema of extremities.
- Dryness and fissuring of the lips.
- Strawberry like redness and swelling of tongue.
- Acute non-purulent swelling of the lymph nodes.

Myofascial Pain Dysfunction Syndrome

- Pain in the muscle, muscle tenderness of masticatory muscle.
- Clicking or popping noise in temporomandibular joint and limitation of jaw motion.

Nance-Horan Syndrome

- X-linked congenital cataract.
- Supernumerary teeth.
- Incisor teeth can resemble Hutchinson's incisor.

Nager's Syndrome

- It is also called as '*acrofacial dysostosis*'.
- *Facial features*—there is hypoplasia of malar bones, anti-mongoloid obliquity or palpebral fissures. Also absent eyelashes, deformed ears, defective hearing, syndactyly and abnormalities of humerus and radius.
- *Oral features*—orally, there is cleft palate, micrognathism and malocclusion.

Neck-tongue Syndrome

- Unilateral upper nuchal or occipital pain, with or without numbness in the area.
- Simultaneous numbness of the tongue on that side.

Noonan's Syndrome

- Congenital heart disease, chest deformity and mental retardation.
- Short stature, facial bone anomalies and cryptorchidism.

Orofacial Digital Syndrome

- It is X-linked condition which is exclusively found in females.
- *Facial features*—it includes frontal bossing, hypoplasia of alar cartilages, broad nasal root, and ocular hypertelorism.
- *Digital malformation*—it includes clinodactyly, syndactyly, brachydactyly and polydactyly.
- *Oral features*
 - Cleft tongue, cleft of alveolar process (mandibular) and cleft lip.
 - Thick fibrous bands in the lower mucobuccal fold eliminating the sulcus.
 - Supernumerary canine and premolars.
 - Malposition teeth.

Oromandibular Limb Hypogenesis Syndrome

- *Eye lesion*—it consist of ocular hypertelorism.
- *Nerve lesion*—it includes cranial nerve palsy.
- *Hypodactyly*—absence of digit of hands and feet.
- *Hypomelia*—hypoplasia of part of limb.
- *Oral features*—there is hypoglossia, microstomia, cleft palate, ankyloglossia, micrognathia and conical shaped mandibular incisors.

Otopalatodigital Syndrome

- It is also called as '*OPD*' syndrome.
- *General*—affected individuals are mentally subnormal, occasionally deaf and there is generalized bone dysplasia.
- *Facial features*—prominent supraorbital ridge, apparent hypertelorism and frontal bossing.
- *Oral features*—there is cleft palate, micrognathic mandible and genital angle is obtuse.

Papillion-lefevre Syndrome

- Juvenile periodontitis, gingival ulcers, inflammatory gingival enlargement and formation of pockets.
- Palmar plantar hyperkeratosis.
- Generalized hyperhydrosis and dirty colored skin.

Para-trigeminal Syndrome

- Severe headache or pain in the area of trigeminal distribution with signs of ocular and sympathetic paralysis.

Patau's Syndrome

- It is also called as '*trisomy 13 syndrome*'.

- The affected individuals may show polydactyly and heart anomalies.
- *Facial features*—it includes microcephaly, microphthalmia, ocular hypertelorism and deafness.
- *Oral features*—it includes cleft lip and cleft palate.
- Polydactyly and heart anomalies.

Parry-Romberg Syndrome

- It is also called as '*progressive hemifacial atrophy*' characterized by unilateral atrophy of face.
- There is also atrophy of skin, subcutaneous tissue, muscle and bone.
- Loss of hair and vitiligo can be seen.
- Contralateral jacksonian epilepsy and trigeminal neuralgia also occur.
- *Oral features*—atrophy of half of the tongue and retarded dental eruption on the involved side.

Peutz-Jeghers Syndrome

- It is also called as '*hereditary intestinal polyposis syndrome*'.
- There is familial generalized intestinal polyposis.
- Pigmentation of face, oral cavity and sometimes on hands and feet.
- There may gastric, duodenal and colonic adenocarcinoma.

Pfeiffer Syndrome

- *Skeletal deformities*—it includes craniosynostosis with turri-brachycephaly, broad thumbs and halluces.
- *Facial deformities*—midface hypoplasia, shallow orbit, hypertelorism, proptosis and antimongoloid obliquity.
- *Oral features*—it includes maxillary underdevelopment resulting in mandibular prognathism, high arched palate and bifid uvula.

Pierre Robin Syndrome

- There is U-shaped cleft palate, micrognathia and glossoptosis.
- Occasionally hydrocephaly or microcephaly can be seen.

Plummer-Vinson Syndrome

- Cracks or fissures at the corner of mouth (angular cheilitis).
- Dysphagia due to esophageal webs.
- Atrophy of filiform papillae and koilonychia.

Portsmouth Syndrome

- Normal ADP-induced platelet aggregation but abnormal or absent collagen induced aggregation.
- It is associated with thrombocytopenia purpura.

Popliteal Pterygium Syndrome

- Congenital lip pit.
- Popliteal webbing (pterygia).
- Cleft lip/cleft palate.
- Genital abnormalities.
- Congenital bands connecting the upper and lower jaws (syngnathia).

Proteus Syndrome

- *Skull features*—hemihypertrophy of the skull, seldom of other bones of the facial skeleton.
- *Partial gigantism*—partial gigantism especially of hand and/or feet, asymmetric limb overgrowth and length discrepancy.
- *Scoliosis*—scoliosis of the vertebral column.
- *Other features*—hyperostosis, various tumors, lipoma, lymphangioma, connective-tissue nevi, and vascular malformations
- *Teeth features*—late mixed dentition with an unusual asymmetric dental development and eruption and crowding with loss of space in teeth.

Raeder's Syndrome

- It is also called as '*paratrigeminal syndrome*'.
- Headache or pain in the area of trigeminal distribution.
- Ocular sympathetic paralysis.

Ramon's Syndrome

- Hypertrichosis, epilepsy and mental retardation.
- Gingival fibromatosis and cherubism.

Raley-day Syndrome

- Congenital absence of tongue papillae.
- Vasomotor dysfunction, loss of reflexes and feeding problems.
- Lack of pain and taste sensation.

Ramsay Hunt Syndrome

- Zoster infection of geniculate ganglion with involvement of external ear and oral mucosa.
- Facial paralysis, pain of external auditory meatus and pinna of the ear.
- Vesicular eruptions in the oral cavity and oropharynx with hoarseness, tinnitus, and vertigo.

Reiter's Syndrome

- *Urethritis*—urethral discharge is associated with itching and burning sensation.
- *Arthritis*—it often bilaterally symmetrical and usually polyarticular.
- *Conjunctivitis*—it is often mild.
- *Mucocutaneous lesions*—they are similar to blennorrhagia and consist of red or yellow keratotic macules or papules which eventually desquamate.
- *Oral features*—there is recurrent oral ulcerations.

Rieger's Syndrome

- *Facial*—there is broad nasal root, protruding lower lip.
- *Ocular findings*—it includes blue sclera, aniridia glaucoma and microcornea hypoplasia.
- *Oral features*—it includes hypodontia, enamel hypoplasia, maxillary hypoplasia, malformed anterior teeth and microdontia.

Rutherford's Syndrome

- Congenital enlargement of the gingivae and altered eruption of teeth.
- There is corneal dystrophy, conjunctivitis and mucocutaneous lesion.

Rubinstein-Taybi Syndrome

- It is associated with *talon's* cusp.
- Developmental retardation, broad thumb and great toes.
- Delayed or incomplete descent of testes in males.

Sanfilippo's Syndrome

- Severe CNS defect, mild somatic disturbance.
- Enamel hypoplasia and excessive dentinogenesis with obliteration of pulp chambers.

Saethre-Chotzen Syndrome

- It is an autosomal dominant trait characterized by short stature and mild mental retardation.
- *Facial deformities*—it includes ocular hypertelorism, ptosis of eyelids, deviated nasal septum and mild conductive deafness.
- *Other deformities*—finger exhibits cutaneous syndactyly and occasional renal abnormalities can be seen.
- *Oral features*—it includes prognathism, high arched palate and resultant malocclusion.

Scheie's Syndrome

- Stiff joints, corneal clouding, aortic regurgitation and normal intelligence.

Scalded Skin Syndrome

- Mild bullous drug eruption resulting in peeling off large sheet of skin giving the appearance of scalding burn.

Scheuthauer-Marie-Sainton Syndrome

- It is also called as '*cleidocranial dysplasia*'.
- Open fontanelle of skull and partial or complete absence of clavicles.
- Underdeveloped maxilla, multiple impacted or unerupted, permanent or supernumerary teeth.

Senear-Usher Syndrome

- Bulla and vesicle on skin and oral mucosa.
- Nikolsky's sign is positive.
- This lesion is terminated in pemphigus vulgaris or foliaceus.

Sjögren's Syndrome

- *Primary*—keratoconjunctivitis sicca and xerostomia.
- *Secondary*
 - Keratoconjunctivitis sicca and xerostomia.
 - Lupus erythematosus, polyarteritis nodosa, scleroderma and rheumatoid arthritis.

Stevens-Johnson Syndrome

- *Oral mucus membrane lesions*—severe form of erythema multiforme.

- *Eye lesions*—photophobia, conjunctivitis, corneal ulceration.
- *Genital lesions*—nonspecific urethritis, balanitis, or vaginal ulcers.

Sturge-Weber Syndrome

- Angiomatosis of face (nevus flammeus) and leptomeningeal angiomas.
- There is also intracranial calcifications and contralateral hemiplegia.
- Massive growth of gingiva and asymmetrical jaw growth and tooth eruption sequence.

Sweat Retention Syndrome

- Extravasation of sweat or saliva in the tissue with subsequent inflammation.
- Keratin plugs formation in sweat glands and accessory salivary glands.

Sweet Syndrome

- Fever.
- Relative increase of neutrophils in the peripheral blood.
- Skin lesion such as tender erythematous plaque, nodules, vesicle and pustules on the face and extremities.
- Dense dermal infiltrate with mature neutrophils.
- Intraoral ulceration is also seen.

SUNCT Syndrome

- S—short acting
- U—unilateral
- N—neuralgiform headache
- C—conjunctival infection
- T—tearing

Treacher Collins Syndrome

- It is also called as '*mandibulofacial dysostosis*' and is autosomal dominant.
- *Facial features*
 - Downward sloping of palpebral fissures (antimongoloid obliquity), under-developed cheek bone, receding chin and malformation of external ear.
 - Hypoplasia of facial bone, especially of malar bone and mandible.
 - Blind fistulae between angle of mouth and angle of ear.
 - Facial clefts and skeletal deformities.
- *Oral features*—fish like mouth, macrostomia, high arched palate, malocclusion, pronounced concavity of the under-surface of the mandible and an obtuse mandibular angle.

Trotter's Syndrome

- Carcinoma of nasopharynx often producing trigeminal neuralgia like pain in the mandible, tongue and side of the head.
- There is also middle ear deafness.

Trichodental Syndrome

- Inherited dominant condition and is characterized by fine short hair.

- Thinning of lateral end of eyebrows.
- Hypodontia and conical teeth.

Trichodentoosseous Syndrome

- It is transmitted as an autosomal dominant trait.
- *Hair and nail deformities*—it includes kinky hair and nails show white band and are brittle.
- *Oral features*—it includes hypomaturation type of amelogenesis imperfecta, enamel hypoplasia, unerupted teeth and taurodontism. The mandibular angle is obtuse and the jaw is square.

Trichonychodental Syndrome

- It is autosomal dominant trait.
- Fine curly hair and thin dysplastic nails.
- *Oral features*—taurodontism and developmental defect of enamel and dentin.

Turner's Syndrome

- Short stature, cabitus vulgus, webbed neck, renal disorders and sexual infantilism.
- *Oral features*—it includes micrognathia, high arched palate and corners of mouth appear pulled down. There is premature eruption of teeth.

Tuberous Sclerosis Syndrome

- *Oral*—gingival lesions and enamel hypoplasia.
- *Skin*—adenoma sebaceum.
- *Associated anomalies*—epilepsy, mental retardation and hamartomas of brain, heart and kidney.
- *Associated malignancy*—astrocytoma and glioblastoma.

van der Woude's Syndrome

- Occurrence of pits of lower lip
- Presence of cleft lip and cleft palate.

Van Buchem's Syndrome

- Excessive deposition of endosteal bone throughout the skeleton.
- Facial swelling and occasional facial paralysis.

Velocardiofacial Syndrome

- *Synonym*—it is also called as Shprintzen Syndrome, Craniofacial Syndrome, or Conotruncal Anomaly Face Syndrome. The name Velocardiofacial Syndrome comes from the Latin words “velum” meaning palate, “cardia” meaning heart, and “facies” means facial features.
- *Cleft palate*—cleft palate (incomplete closure of the roof of the mouth).
- *Heart defect*—multiple abnormalities of the heart including Ventricular septal defect (VSD), pulmonary atresia, tetralogy of Fallot, truncus arteriosus, interrupted or right-sided aortic arch and transposition of the great arteries.

- *Facial features*—there is long face with prominent upper jaw, flattening of the cheeks and underdeveloped lower jaw. There is also prominent nose with narrow nasal passages and thin upper lip with a down-slanted mouth.
- *Other features*—minor learning problems, speech and feeding problems, bluish color below the eyes can also be present.

von Recklinghausen's Neurofibromatosis

- *Oral lesions*—intraoral neurofibromas leading to macroglossia.
- *Skin lesion*—café au lait spots, giant nevi and multiple neurofibromas.
- *Malignancy*—malignant neurilemmoma and pheochromocytomas.
- *Neurological*—CNS tumor, mental retardation.

von Hippel's Syndrome

- Hemangioblastomas in retina and cerebellum.
- Pancreatic and renal cyst, renal adenomas and hepatic hemangioma.
- Multiple endocrine neoplasms.

Whistling Face Syndrome

- It is also called as ‘*craniocarpotarsal dysplasia*’.
- It is characterized by sunken eyes, true ocular hypertelorism and antimongoloid obliquity of palpebral fissures.
- It also includes small nose, microstomia, high skull and protruding lips as seen during whistling.
- The palate is high arched and mandible is small and retrognathic.
- There is presence of a fibrous band demarcated by two grooves extending from the midline of the lower lip to the chin, often presenting in ‘H’ or ‘V’ shaped.

Weber Cockayne Syndrome

- It is localized form of epidermolysis bullosa.
- Bullae develop on hand and feet and are related to frictional trauma.
- There is no scarring upon healing.

XXXXY Syndrome

- There is hypoplastic midface, short stature, mental retardation, speckled eyes and hypertelorism.
- *Oral features*—it includes taurodontism and bifid uvula.

Zimmerman-Laband Syndrome

- Gingival fibromatosis and ear, nose and nail defect.
- Hepatosplenomegaly and hyperextensible joint.

Zinsser's Syndrome

- Oral leukoplakia, dystrophic nails and hyperpigmentation of skin.
- Pancytopenia and aplastic anemia.

3

Glossary

Aberration—it is a variation from the normal form or course.

Aberrancy—it is defined as that situation in which a tissue develops at a site where it is not normally found.

Ablation—it is removal of a part by excision or amputation.

Abnormal—it is not normal, deviating in some from the usual structure, position or state.

Abocclusion—it is a condition where the maxillary and mandibular teeth are not in contact.

Abrasion—it is the wearing away of a structure or substance by mechanical means such as scrubbing or grinding.

Abrasive—it is a substance which contains an abrasive which tends to erode the surface.

Abfraction—loss of tooth surface at the cervical areas of teeth, caused by tensile and compressive forces during tooth flexure; cervical erosive lesions that can not be attributed to any particular cause.

Abscess—an abscess is a localized collection of pus surrounded by an area of inflamed tissue in which hyperemia and infiltration of leukocytes is marked.

Actinic keratosis—it is a premalignant squamous cell lesion resulting from long-term exposure to solar radiation and may be found on the vermillion border of lip as well as other sun exposed skin surfaces.

Actinic elastosis—it is a lesion on the labial mucosa exposed to sun. A white area of atrophic epithelium develops with underlying scarring of the lamina propria.

Actinic cheilitis—when this atrophic tissue abrades to ulcer, it is called as actinic cheilitis.

Acanthosis—this condition is characterized by widening and thickening of stratum spinosum.

Acantholysis—it is the pathological separation of epidermal or epithelial cells by breakdown of desmosomes in stratum spinosum (seen in pemphigus).

Acquired—relating to something not of genetic origin but resulting from outside influence.

Acrocephalic—it is a highly arched or pointed skull.

Acute—having severe symptoms and a short course.

Adduction—drawing in towards the center or to median line, as opposed to abduction.

Adenomatosis oris—it is the swelling of the mucous glands of the lips with no inflammation or secretion.

Adrenodontia—it is a morphological indication of over activity of adrenal glands characterized by large pointed canines and teeth with occlusal surfaces showing brown discoloration.

Aerodontia—it is that branch of dentistry concerned with the care and treatment of dental conditions caused by high altitude flying.

Afferent nerve—it refers to any nerve transmitting impulse from the periphery to the center.

Ageusia—it is the loss or absence of sense of taste.

Aglossostomia—it is the congenital absence of the tongue and of mouth opening.

- Agranulocytosis**—a marked decrease in the number of granulocytes, particularly neutrophils.
- Allograft**—a graft using material not derived from a donor or from animal sources, e.g. synthetic resins, stainless steel alloy.
- Allergen**—a substance capable of inducing hypersensitivity or an allergic reaction.
- Allergy**—it is hypersensitivity to any normally harmless substance resulting in an exaggerated or abnormal reaction.
- Allograft**—it is a graft derived from a donor of the same species but genetically dissimilar.
- Amniocentesis**—it is diagnostic procedure in which a small amount of amniotic fluid is withdrawn from amniotic sac, a membrane surrounding the fetus in uterus, to detect fetal defects.
- Amalgam tattoo**—oral soft tissue discolorations due to amalgam; most common pigmentation of the oral cavity.
- Amelogenesis**—the formation of the enamel portion of the tooth.
- Analgesia**—it is relief from pain or insensibility to pain.
- Analogous**—having similar properties.
- Anesthesia**—it is the general loss of all sensations or feelings.
- Anemia**—it is an abnormal reduction in the number of circulating red blood cells, the quantity of hemoglobin and the volume of packed red cells in a given unit of blood.
- Anaphylaxis**—it is an antigen-antibody reaction produced by the parenteral injection of an antigen causing hypersensitivity.
- Anastomosis**—it is a communication between two vessels.
- Anachoresis**—if the bacteria circulating in the bloodstream settle in areas of inflammation or of lowered resistance in the pulp and produce pulpitis, abscess or necrosis, the phenomenon is referred to as anachoresis.
- Anomaly**—deviation or irregularity as compared with the normal.
- Anorexia**—it is the lack of appetite.
- Anomalad**—it is a malformation together with its subsequently derived structural changes; the primary defect setting off a series of secondary or even tertiary events resulting in multiple anomalies
- Anosmia**—it is the absence of sense of smell.
- Antagonist**—it is any tissue that acts against or in opposition to another tissue.
- Anaplasia**—it is the reversion of the same type of cells from a more highly differentiated to a less highly differentiated type.
- Antibody**—it is any one of the class of substances produced in the body as a reaction to a specific antigen and with which, it reacts in some observable way to produce a specific effect such as inactivation, agglutination, and/or flocculation.
- Antibiotics**—these are substances produced by microorganisms which suppress the growth or kill other microorganisms at a very low concentration.
- Antidote**—it is an agent used to counteract or prevent the action of poisons.
- Antigen**—it is any substance that when introduced into the body, excites the formation of specific antibodies.
- Angioma**—a tumor made up of blood or lymph vessels.
- Ankyloglossia**—extensive adhesion of the tongue to the floor of the mouth or the lingual aspect of the anterior portion of the mandible caused by a short lingual frenum.
- Apertognathia (open bite)**—a condition in which the anterior or the posterior teeth of the mandible can not be brought into occlusion with antagonist teeth of maxilla.
- Aponeuroses**—these are collagenous sheets or ribbons that resemble flat, broad tendons. It may cover the surface of the muscle and assist in attaching superficial muscles or separate the structures.
- Aplasia**—absence of an organ or organ's part due to failure of development of the embryonic tissue of origin.
- Arteriosclerosis**—a condition characterized by loss of elasticity and thickening of artery walls.
- Atrophy**—it is a reduction in size of tissue or of an organ due to decrease in the size or number of its constituent cells.
- Atresia**—it is the congenital occlusion or absence of one or two major salivary gland ducts.

Atypical—irregular, not conformable to the type.

Attrition—it is the physiologic wearing away of tooth material as a result of tooth to tooth contact.

Auscultation—listening to the sound produced within the body with the help of a stethoscope.

Autogenous—it is produced within the body itself. It is self-generated.

Autograft—it is a graft taken from one of the patient's body and transplanted to another part in the same individual.

Autoantibody—an antibody that reacts against an antigenic constituent of the person's own tissues.

Autoimmune disease—a disease characterized by tissue injury caused by a humoral or cell-mediated immune response against constituents of the body's own tissues.

Autoimmunity—immune-mediated destruction of the body's own cells and tissues; immunity against self.

Autosomes—the non-sex chromosomes that are identical for men and women.

Autoinoculation—to inoculate with a pathogen such as a virus from one's own body.

Bacteremia—it refers to the circulation of bacteria in the blood.

Bacteria—these are microscopic unicellular vegetative organisms having a single chromosome, no nuclear envelope and a rigid cell wall. They may be seen as rods, cocci or filaments and divide by binary fission.

Bacteriostatic—it is any agent that inhibits the growth and multiplication of bacteria.

Baelz's disease—it is a disease characterized by the presence of painless papules on the labial mucous membrane (Cheilitis glandularis-superficial suppurative type).

Ballooning degeneration—it is characterized by the isolation of a cell from its neighbors, especially in the lower layers of the epidermis, the withdrawing of its prickles after intra-cytoplasmic edema and vacuolization and the mitotic division of its nucleus so as to form multinucleated giant cells.

Bay cyst—apical cyst which have a direct connection with apical foramen have been termed as 'bay cyst'.

Bednar' aphthae—two ulcers appearing symmetrically one on either side of the midline of the hard palate in infants, thought to be caused by the nipple or by thumb sucking or sucking hard object.

Benign—not malignant; favorable for recovery.

Bicameral abscess—it is an abscess which contains two chambers.

Biopsy—it is the gross and microscopic examination of tissue or cells removed from living patients for the purpose of diagnosis or prognosis of the disease or the confirmation of the normal condition.

Blanching—to take the color out of and make white.

Bleb—it is a bulla or other skin blister filled with blood or serous fluid.

Blind abscess—it is the one having no fistulous tracts.

Blister—it is a vesicle caused by localized accumulation of fluid beneath the skin.

Blood—it is the red fluid in the vessels of the circulating system which conveys oxygen and nutritive materials to the tissue and removes carbon dioxide and waste matter.

Blood pressure—it is the pressure exerted by the blood on the artery walls and is dependent on the force of heart action, the elasticity of the vessel walls, capillary resistance and the volume and viscosity of blood.

Blood transfusion—the intravenous administration of blood to help replenish excess blood loss due to hemorrhage or other wise, is known as blood transfusion.

Boil—it is a localized skin abscess usually at the site of a hair follicle.

Bosselated—having a knob like protrusion or bosses.

Bowen's disease—it is a localized intra-epidermoid carcinoma that may progress to invasive carcinoma over many years.

Bradycardia—it is an abnormal slowness of the heart and pulse rate.

Bradyglossia—it is an abnormal slowness of speech, due to difficulty in tongue movements.

Bradypnea—it is an abnormal slowness of respiration.

Bruise—it is a superficial injury, caused by a blow with no laceration but with discoloration of the skin and subcutaneous tissue produced by an accumulation of blood.

Bruxism—it can be defined as the involuntary, unconscious, and excessive grinding, tapping or clenching of teeth *or* it is defined as non-functional grinding or gnashing of the teeth, usually during sleep.

Buccal bifurcation cyst—a cyst of uncertain origin found primarily on the distal or facial aspect of a vital mandibular third molar, consisting of intensely inflamed connective tissue and epithelial lining.

Bullae—it is an elevated blister like lesion containing clear fluid and is bigger than 1cm in diameter.

Burrows—these are short, linear, straight or sinuous lines in the skin.

Burn—it is the injury resulting from the application of excessive heat, electric current, friction and caustics to skin or mucous membrane.

Carcinogenesis—carcinogenesis or oncogenesis or tumorigenesis means induction of a tumor agent which can induce tumor. The tumor agents are called as carcinogens.

Carabelli's cusp—it is an accessory lingual cusp located on mesiopalatine cusp of maxillary second primary molars and 1st, 2nd and 3rd permanent molars.

Capsule—compressed fibrous connective tissue around a benign neoplasm separating it from surrounding tissues.

Carcinoma—a malignant growth made up of epithelial cells that are capable of infiltration and metastasis.

Caries—demineralization of inorganic and dissolution of organic part of the tooth surface caused by bacteria.

Carcinoma in situ—it is a histopathological diagnosis defined as a proliferation of basal epithelial cells from the basement membrane to the surface, with almost all of the cells manifesting cytologic atypia. Immediate maturation into a superficial keratin layer is possible, but no invasion into the underlying connective tissues can be seen.

Calcareous—relating to or containing calcium or calcium salts; chalky.

Calcification—it is the deposition inorganic tissue of calcium salts causing hardening.

Calcinosis—it is a condition characterized by either localized or generalized deposition of calcium salts in nodules in the soft tissues.

Callus—the mesh of fibrous bony tissue surrounding and uniting the bone ends after fracture. It is later replaced by hard bone.

Camper's line—it is the line extending from the external auditory meatus to a point below the nasal point and is also called as facial line.

Cancellous—having a lattice like spongy structure; applied to bone tissue.

Canker—it is an ulceration especially of the mouth and lips and it is also called as aphthous stomatitis.

Capillary—these are one of the very fine thread like blood vessels connecting the veins and arteries.

Carbuncle—it is a staphylococcal infection of the sweat glands or hair follicles causing inflammation of the surrounding subcutaneous tissue and discharging pus through several openings, finally sloughing away.

Carcinosarcoma—it is a mixed tumor containing characteristics of both carcinoma and sarcoma.

Cariology—it is the scientific study of dental caries, its causes, prevention and treatment.

Carrier—the individual who continues to harbor infectious agent either following recovery from the illness it induced.

Cartilage—it is a form of elastic, nonvascular connective tissue attached to articular bone surfaces and also forming some parts of the skeleton.

Catabolism—it is the process of breakdown of complex compounds by the body.

Catarrh—it is the inflammation of the mucous membranes, especially those of nose and throat, with discharge of mucus.

Causalgia—it is a burning sensation arising after trauma to a sensory nerve.

Cellulitis—cellulitis may be defined as a non-suppurative inflammation of the subcutaneous tissue extending along the connective tissue planes and across the intercellular spaces.

Cell—it is one of the minute masses of protoplasm, containing a nucleus which forms the basis of all animal and plant structure.

Cementicle—it is a small calcareous body developing in the periodontal membrane.

Cell mediated immunity—it is the type of immunity in which the predominant role is played by T-lymphocytes.

Central—in oral pathology, it is the lesion occurring within bone.

Centromere—the constricted portion of the chromosome that divides the short arms from the long arms.

Chief complaint—it is the patient's response to the dentist's question.

Cheilitis—it is the inflammation of lip.

Chemoprophylaxis—it is the use of chemical drugs in the prevention of disease.

Chemotherapy—it is the treatment of a disease by chemicals which affect pathogenic organisms without harming the patient or it is the treatment of malignant neoplasia by chemical means.

Cheesy—lesion's texture is similar to curd of cheese.

Chemotaxis—taxis or movement in response to chemical stimulation.

Chromatin—a general term used to refer to the material (DNA) that forms the chromosomes.

Chronic—persisting over a long time; when applied to a disease, chronic means that there has been little change or extremely slow progression over a long period.

Chills—it is cold sensation with shivering, often characteristic of onset of fever.

Chloroma—it is a condition characterized by multiple myeloid tumors of greenish color, affecting particularly the face and skull, and associated with blood picture of leukemia.

Choriostoma—it refers to excessive amount of normal tissue that is present in abnormal location.

Chondromalacia—it is a condition characterized by abnormal softness of the cartilage.

Ciliated—having hair like processes or fringe of hair.

Circulation—it is the movement or flow in a circle, retracing its course repeatedly, applied especially to the flow of blood through the body.

Cleft lip—it is a birth defect that results in a unilateral or bilateral opening in the upper lip between the mouth and the nose.

Cleft palate—cleft palate is a birth defect characterized by an opening in the roof of the mouth caused by lack of tissue development.

Coagulation—when blood is shed, it loses its fluidity in few minutes and sets into a semisolid jelly. This is called as coagulation or clotting.

Cold abscess—it is a slow developing tuberculous abscess generally about a bone or joint and with little inflammation.

Collar stud abscess—it is a superficial abscess connected by a sinus tract to a larger deep abscess.

Complement system—this consists of a group of serum proteins which by series of reactions produce and release by products whose functions are to initiate an inflammatory reaction, to regulate and enhance phagocytic function and attack the bacterial cell membrane.

Congenital—present at or before birth but not necessarily inherited.

Coalesce—it is a term used to denote to fusion or union of separated parts.

Consanguinity—blood relationship. In genetics, the term is generally used to describe marriages among close relatives.

Corrugated—having a surface that appears wrinkled.

Cotton wool—confluent radiopacities.

Coarctation—it is narrowing or constriction, applied especially to blood vessels.

Col—it is a depression in an interdental papilla between the two peaks, one on each side of the contact area.

Coma—it is a state of complete unconsciousness from which a patient can not be aroused, even by determined external stimulation.

Commensal—it is an organism that lives on or within another organism, to its own advantage and without being detrimental to the host.

Commissure—it is the point of union between similar parts or bodies.

Concretion—it refers to any hardened or solidified mass in the tissue.

Counter irritation—it refers to the deliberate production of superficial irritation in order to mask or relieve an existing irritation or pain.

Concrescence—it is a form of fusion that occurs after the root and other major parts of the involved teeth are formed or when the roots of two or more teeth are united by cementum, below the cemento-enamel junction.

Cranio-malacia—it refers to a condition characterized by softness of bones of the skull, usually seen in infants.

Crater—it is a localized depression, usually circular, with raised edge or rim.

Crust—dry products of exudation from lesions occurring on skin and lips.

Crepitations—it refers to a crackling noise occurring in the joint when affected by certain disease.

Cryosurgery—it is the use of extreme cold for surgical destruction of tissue.

Cryotherapy—it is the treatment of disease with use of extreme cold.

Cryptogenic leukoplakia—in a small proportion of cases of leukoplakia, no underlying cause has been found. Such lesions are termed as idiopathic or cryptogenic leukoplakia.

Culture—it is the growth of microorganisms in an artificial medium.

Curettage—it refers to the removal of foreign matter from the walls of a bony cavity or from the root surface.

Cyst—cyst is a pathological cavity which may or may not be lined by epithelium and consists of fluid, semi-fluid or gaseous content (but not by pus) and surrounded by connective tissue capsule. True cyst is a pathologic cavity always lined by epithelium usually containing fluid or semi-solid material.

Cytology—is the scientific study of cell.

Cytopathic—pertaining to or characterized by pathologic changes in cells.

Cyanosis—it is the bluish discoloration of the skin and mucous membranes, often due to deficient oxygenation of the blood.

Dental kinesiology—it is the study of motion and function of jaws and oral musculature; the accompanying neurological, vascular and other supporting system network and the impact of those muscle functions and neurological dynamics have on dental and systemic health.

Developmental anomalies—malformation or defects resulting from disturbance of growth and development are known as developmental anomalies.

Dens in dente—it is also called as dens invaginatus. Infolding of the outer surface of the tooth into interior. It is a developmental variation which is thought to arise as a result of invagination in the surface of tooth crown before calcification occurs.

Dens evaginatus—dens evaginatus is a developmental condition that appears clinically as an accessory cusp or globules of enamel on occlusal surface between buccal and lingual cusp of premolars.

Debridement—it is the removal of dead tissue and foreign matter from a wound.

Degeneration—it refers to the gradual deterioration of tissue with loss of function and chemical changes within the tissue.

Dentistry—it is a branch of medicine concerned with oral and dental diseases and their prevention and treatment and with oral prosthesis.

Dentoalveolar abscess—an abscess that forms at the end of the tooth root.

Desmosomes—the term desmosomes refers to the structures forming the site of contact between adjacent cells, especially epithelial cells.

Desquamation—it refers to the peeling off of the outer layer of epithelium.

Dental fluorosis—a condition of enamel hypoplasia characterized by white chalky spots or brown staining and pitting of teeth due to an increased level of fluoride; affecting enamel matrix formation and calcification by impairment of ameloblastic function.

Dentigerous cyst—an odontogenic cyst that surrounds the crown of an impacted tooth; caused by fluid accumulation between the reduced enamel epithelium and enamel surface, resulting in a cyst.

Deoxyribonucleic acid (DNA)—a substance composed of a double chain of polynucleotide; both chains coiled around a central axis form a double helix. DNA is the basic genetic code or template for amino acid formation.

Dermoid cyst—a cyst of midline of the upper neck or the anterior floor of the mouth of young patients, derived from remnants of embryonic skin; consisting of a lumen lined by a keratinizing stratified squamous epithelium and containing one or more skin appendages such as hair, sweat or sebaceous glands.

Diffuse—used in the description of a lesion; when borders of the lesion are not well defined and it is not possible to detect the exact parameters of the lesion, then this term is used.

Diploid—having two sets of chromosomes; the normal constitution of somatic cells.

Diagnosis—it is the determination of the nature or cause of the disease.

Differential diagnosis—the list of similar clinical picture, according to probable identity of condition at hand, is the differential diagnosis.

Dimorphic anemia—it is a condition in iron deficiency and folic acid deficiency anemia can occur concomitantly.

Diploe—the spongy layer of bone position between the inner and outer layers of compact bone.

Diverticuli—they are small pouches or out pocket of the ductal system of one of the major salivary glands.

Disinfection—this is the process by which pathogenic microorganisms are removed from the surface, without removing bacterial spores.

Dilacerations—it refers to angulations or sharp bends or curves in the root and crown of the teeth.

Disease—it is the departure from the average anatomical structure or is an abnormal degree of failure of physiological function or some reduction in psychological efficiency, due to either adversity in the genetic endowment of the individual or misuse of his free will or to adverse factors in the environment in which he lives or some combination of these factors or it is defined as loss of ease.

Distomolar—found in the molar region frequently located distal to 3rd molar.

Discrete—separate. Composed of separate parts, not joined or blended.

Dislocation—it is the displacement of any part from its normal position, especially in the cases of bone and joints.

Direct fracture—it refers to the fracture that occurs at the site of blow.

DLE (discoid lupus erythematosus)—it is a circumscribed slightly elevated white patch that may be surrounded by a red telangiectic halo.

Dose—it is one measured portion of any medicine which is to be taken one at a time.

Dominant—in genetics, a trait or characteristic that is manifested when it is carried by only one of a pair of homologous chromosomes.

Dorsal—directed towards or situated on the back surface (opposite of ventral).

Dry abscess—it is an abscess that disperses without bursting or coming to a head.

Drainage—it is the gradual removal of fluid from a cavity or wound.

Dressing—it is a medicament used to promote wound healing or as a covering for a wound, used for protection or to assist healing.

Drug—it is any medicinal substance.

Drug addiction—it is state of periodic or chronic intoxication produced by the repeated consumption of drug and is harmful to individual and to society.

Drug habituation—it is the condition resulting from repeated consumption of drug, in which there is psychological and emotional dependency on the drug.

Drug dependence—a state psychic and sometimes also physical resulting from the interaction between a living organism and a drug, characterized by behavioral and other response that always includes a compulsion to the drug on a continues or period basis in order to experience its psychic effect and sometimes to avoid the discomfort of its absence.

Drug abuse/misuse—it is the improper or excessive use of therapeutic drugs even in the absence of addiction.

Dyskinesia—it is defined as an impairment of voluntary motions, causing movements that are incomplete or only partial.

Dysesthesia—it refers to the impairment of feeling or sensations; a condition in which a normal stimulus produces disagreeable sensations.

Dyskeratosis—this lesion shows abnormal orientation in development of epithelial cells.

Dysodontiasis—it refers to the painful, difficult or delayed eruption of the teeth.

Dysostosis—it refers to the congenital defective bone formation.

Dysphagia—an experience of having great difficulty in swallowing.

Dystrophic calcification—pathologic calcification that occurs in degenerating and dead tissue.

Dysplasia—it refers to the abnormal formation or development.

Dyspnea—it is a shortness of breath.

Ecchymosis—it refers to the diffuse extravasation of blood into the tissues. Larger purpuric lesions are called as ecchymoses.

Ectoderm—the outermost of the three primary germ layers of the embryo, from which are developed the epidermis, the external sense organs and the oral and anal mucous membranes.

Edema—it is accumulation of excess fluid in the intercellular tissue spaces or body cavities.

Electrocautery—it is cauterization by low voltage current producing burn like tissue repair, but with no control over the extent or quality of tissue destruction.

Electrodesiccation—it refers to the deeply penetrating tissue dehydration produced by the insertion of electrodes into the tissue.

Empirical therapy—with serious infections, it is often necessary to begin antibiotic therapy before culture result is available, this is called as empirical therapy which is directed towards organisms which are most likely to have caused that infection.

Embedded teeth—those teeth which are unerupted usually because of lack of eruptive force.

Embolism—it refers to the sudden blockage of blood vessels by a clot or other obstruction within the blood stream, causing failure of circulation.

Empyema—it is the accumulation of pus in a body cavity or a hollow organ.

Enamel pearls, nodules, or droplets—pearls or droplets described as small buttons or nodules of enamel usually about 1 mm or 2 mm in diameter that form on the root or at the bifurcation of multi-rooted teeth.

Embryonic—pertaining to the earliest stage of development of an organism.

Emigration—the passage of white blood cells through the endothelium and walls of small blood vessels.

Endotoxin—they are heat stable phospholipid-polysaccharide-protein complex contained as a structural part of the cell of many Gram-negative bacteria and released by disintegration of the cells.

Enanthema—it is an eruption occurring on a mucous surface or on any surface within the body as opposed to exanthema.

Endemic—prevalent in a particular region.

Endosteal—it is within the bone.

Endothelium—it refers to the membrane lining the heart and blood vessels.

Engorgement—it refers to the excess of blood in any part of the body or it is the localized congestion or distension.

Enostosis—it is a localized morbid bone growth arising within the bone cavity.

Enucleate—the word enucleate means to remove an organ or part, or a circumscribed, space filling lesion entirely, i.e. from its outer sheath or covering.

Endodermal—pertaining to the innermost of the three primitive germ layers of an embryo. Endodermal structures include the epithelium pharynx, respiratory tract (except the nose) and digestive tract.

Epidemic—affecting large number of people within an area or region.

Epidemiology—it is that branch of science concerned with the study of a disease or condition through its frequency and distribution.

Epithelium—it is a thin cellular layer covering or lining the organs and tissues of the body.

Eponym—the name of an organ, syndrome, disease, etc. that contains or is derived from a proper name.

Epulis—any tumor of the gums; more especially either a fibrous or a giant cell tumor.

Eruption—the act of appearing, or pushing through, as of teeth coming through the gums or a visible skin lesion occurring in disease.

Erythema—it is the redness in the skin either diffuse or patchy, caused by congestion of the subcutaneous capillaries.

Erythematous—it is characterized by a redness of the tissue due to engorgement of the capillaries in the region.

Erythroplastic—it is characterized by a reddish appearance. This term implies abnormal tissue proliferation in the reddish area.

Erythrocyte—one of the red cells found in blood which carries oxygen and is produced by the bone marrow.

Erythroplakia—the term is applied to any area of reddened velvety textured mucosa that can not be identified on the basis of clinical and histopathological examination as a cause of inflammation or any other disease process.

Erythrodontia—there is deposition of porphyrins in dentin and to a lesser extent in the enamel which imparts red or brown color to the deciduous and permanent teeth and known as erythrodontia.

Erosion—it is a shallow crater in the epithelial surface that appears on clinical examination as a very shallow erythematous area with only superficial changes or a moist red lesion often caused by rupture in vesicles and bullae as well as trauma.

Erosion (teeth)—it is loss of tooth substance due to chemical process that does not involve bacterial activity.

Erythroplasia—these are painless erythematous eruptions, papular or macular in nature, affecting the mucous membrane.

Eschar—it is a dry slough, the result of burning or due to contact with a corrosive agent.

Etiology—the study or theory of the factors that cause disease and their introduction to the host.

Eversion—a turning outward or a state being turned outwards.

Excrescence—it refers to an abnormal growth protruding from body or plant.

Exacerbation—it refers to an increase in the severity of a disease or any symptoms.

Examination—it refers to investigations carried out for diagnostic purpose.

Exanthema—it is an eruptive fever.

Excoriation—it is the superficial loss of surface skin or a graze.

Excursion—it refers to any movement of a movable part from a resting position during the performance of some functions.

Exfoliation—it is the peeling off in layers or in scales.

Exophthalmos—it refers to the abnormal protrusion of the eyeball.

Exophytic—it refers to a word relating to something growing outwards, used for tumor projecting above the normal surface contours or it refers to any pathological growth that project above the normal contours of the oral surface.

Expansile—capable of being extended or expanded.

Expressivity—in genetics, the degree of clinical manifestation of a trait or characteristic.

Exostosis—it is a bony swelling developing on the bone surface or on a tooth root.

Exotoxin—it refers to a toxic secretion of bacterial cells which cause damage in sites distant from the focus of infections or they are heat labile proteins which are secreted by certain bacteria and diffuse readily into surrounding tissue.

Extravasation—it is the escape of fluid from vessels into the surrounding tissue.

Extrinsic—having its origin outside and separated from a body, organ or part.

Exudate—the matter that passes out into adjacent tissues through vessel walls in inflammation.

Facies—the appearance of the face.

Factitial injuries—these are accidentally self-induced injuries on the basis of habits with frequent psychological backgrounds.

Favorable fracture—if the fracture line runs in such a manner that the associated muscle tends to hold the fragments together, the fracture is described as favorable.

Facet—it is a small abraded area on a bone or on tooth surface.

Familial—relating to a family, or affecting several of its members.

Fascia—it is the layer of areolar tissue beneath the skin or the layer of areolar tissue investing the muscles, nerves and other organs.

Fenestrate—to pierce with one or more holes, sometimes used on the walls of bony defect in an attempt to stimulate repair.

Fenestration—it refers to a surgical procedure by which one or more holes are pierced in hard tissue.

Fever—it refers to an abnormal increase in body temperature.

Final diagnosis—it is statement with which precise diagnosis has been made on the basis of all required observation, identification of definitive symptoms and the pathological report and patient response to therapy.

Fibro-cemento-osseous lesions—it is a skeletal disorder in which bone is replaced by fibrous tissue which in turn is replaced by mineralized tissue.

Fissure—it is a linear often crusted, tender, painful defect in continuity of the skin, occurring usually at the mucocutaneous junctions and at sites where there is considerable elasticity of the skin.

Fistula—it is communicating tract between two epithelial surfaces which is lined by granulation tissue which is subsequently epithelialized.

Fibrosis—there is an abnormal formation of fibrous tissue.

Fluctuant—a wave-like motion felt on palpating a cavity with non-rigid walls, especially one containing fluid.

Fluoride mottling—a condition of enamel hypoplasia characterized by white chalky spots or brown staining and pitting of teeth due to an increased level of fluoride affecting enamel matrix formation and calcification by impairment of ameloblastic function.

Focus of infection—it refers to a circumscribed area of tissue, which is infected with exogenous pathogenic microorganisms and which is usually located near a mucous or cutaneous surface.

Focal infection—it refers to metastasis from the focus of infection of organisms or their products that are capable of injuring tissue.

Foramen—a small hole in a bone through which passes either blood vessels or nerves or both.

Focal osteitis—a condition sometimes occurring after tooth extraction, particularly after traumatic extraction, resulting in a dry appearance of the exposed bone in the socket, due to disintegration or loss of the blood clot.

Foreign body granuloma—a reaction to foreign materials that are too large to be ingested by either microphages (PMNs) or macrophages.

Frenal tag—a redundant piece of mucosal tissue that projects from the maxillary labial frenum.

Fusion—it is also called as synodontia. It represents the embryonic union of normally separated tooth germs.

Fulguration—it refers to the superficial tissue dehydration produced by a surgical electrode held slightly away from the tissue, causing sparking.

Galvanism—the production of an electric current caused when two dissimilar metals used as restorations in the mouth come into contact, this can cause discomfort and even pain.

Gangrene—it is the necrosis of tissue due to failure of the arterial blood supply caused by injury or disease.

Gelation—the process of change of a colloid from a sol to a gel.

Gerodontia—it is that branch of dentistry which deals with the care of old people.

Gemination—it refers to the process whereby single tooth germ invaginates resulting in incomplete formation of two teeth that may appear as a bifid crown on a single root.

Genetic heterogeneity—having more than one inheritance pattern.

Ghost teeth—a developmental disturbance of several adjacent teeth in which the enamel and dentin are thin and irregular and fail to adequately mineralize; surrounding soft tissue is hyperplastic and contains focal accumulations of spherical calcifications and odontogenic rests.

Gingivosis—it refers to the any degenerative condition affecting the gingiva.

Gland—an organ that produces secretions.

Glossodynia—it refers to the burning or painful condition of the tongue.

Gomphosis—it is the firm attachment of two bones without a movable joint.

Gorham's disease—in this condition a large portion of bone disappears with any apparent cause.

Granuloma—a tumor composed of granulation tissue.

Granulomatosis—it refers to the development of multiple granuloma.

Granulation tissue—it is the reparative tissue that is formed on the surface of wound having pink, soft, granular appearance showing histologically new small blood vessel and fibroblast.

Green stick bone fracture—it is a fracture in which one side of bone is broken and the other side is bent but intact.

Ground glass—fine radiopaque spots in radiolucent background.

Gustatory—the sense of taste or the act of tasting.

Hamartomas—it is a tumor like malformation of oral tissues, developmental in origin with tissue being native to the site.

Habit—it is a tendency toward an act or an act that has become a repeated performance, relatively fixed, constant, easy to perform and almost automatic.

Hemoglobinopathies—these are a group of hereditary disorders characterized by the presence of structurally abnormal hemoglobin.

Hereditary disease—they are apparent at birth but some may not become evident for years.

Hemoptysis—the presence of blood in the sputum caused by bleeding in the upper respiratory tract or the lungs.

Hemorrhage—it refers to the internal or external loss of blood due to injury or other damage to blood vessels.

Hemidesmosome—it refers to a structure found on the basal surface of an epithelial cell, the attachment site between the cell and the underlying membrane.

Heredity—it refers to the transmission of a characteristic from parent to child or to later generation.

Heterotrophic—it is a term used relating to organisms which require a complex source of carbon for nourishment and growth.

Hematoma—it is large clot resulting from blood released into the tissue from a ruptured or injured blood vessel.

Healing—it is repair and replacement of dead or damaged cells by healthy cells.

Histology—it refers to the study of the anatomy and physiology of tissue and cells using microscopic technique.

Holistic—it refers to an approach to treatment that takes into consideration the whole person, not just the disease or condition.

Homologous—having the same or corresponding structure or position but not necessarily similar in function.

Horner's teeth—incisor teeth with horizontal grooves caused by enamel deficiency.

Hypodontia—it refers to the absence of one or more teeth.

Hypertrophy—it refers to the enlargement caused by an increase in size of cells.

Hydrocyst—it refers to a cyst whose contents are watery in nature.

Hyperplasia—it refers to the enlargement caused by increase in number of cells.

Hypoplasia—it is the failure of full development of an organ or tissue.

Hydrostomia—it refers to a condition characterized by constant dribbling from the mouth.

Hygroma—it refers to a swelling caused by fluid surrounding an inflamed bursa, or distending a sac or cyst.

Hypertension—exceptionally high tension especially abnormally high blood pressure.

Hypnosis—it refers to a sleep or a trance state, especially one induced artificially by verbal suggestions or concentration upon some object.

Hypsodont—having teeth with long crowns and short roots seen in herbivorous animals.

Hydropic degeneration—it refers to replacement of the nuclei of stratum basal by clear space due to edema and degeneration of cells.

Iatrogenic diseases—these are the diseases produced by the action of a doctor or due to medical treatment.

Idiopathic—it is any spontaneous or primary disease with no apparent external cause

Idiosyncrasy—it refers to a reaction to a particular drug in therapeutic doses in a manner not necessarily related to its pharmacological properties.

Impacted teeth—they are those prevented from erupting by some physical barriers in the eruption path.

Immunity—it is the resistance exhibited by the host towards injury caused by microorganisms and their products.

Impermeable—not permitting passage especially of fluids.

Implant—the word implant means to insert into the body or to graft as in plastic surgery.

Infection—it is a clinicopathological entity-involving invasion of the body by pathologic microorganisms and the reaction of tissues to microorganism and their toxins.

Inspection—it refers to an examination of the affected part of the body.

Internal derangement—it can be defined as mal-relationship of the meniscus to the condylar head and articular eminence where an alteration of its attachment allows the meniscus to assume an abnormal position.

Inflammatory collateral cyst—it is a cyst which arises in the periodontium of an erupted tooth as a result of inflammatory process in the periodontal pocket.

Involucrum—small section of necrotic bone may be completely lysed, while a large one may get localized, and get separated and form shell of new bone called involucrum by a bed of granulation tissue or a sheath particularly new bone sheath that forms about sequestration.

Indirect fracture—fracture site distant from where the actual blow takes place, usually seen on contralateral side.

In vitro—within glass referring to observations made in a test tube or culture dish as opposed to *in vivo*.

In vivo—within a living organism.

Indentation—it refers to the condition of being serrated or notched.

Induced—brought on by an outside agent or is artificially produced.

Induration—it refers to the state of being hard or the process of becoming hard.

Inflammation—it is the reaction of living tissue to injury.

Infarction—it is a localized area of ischemic necrosis in an organ or tissue resulting from sudden reduction of either its arterial supply or venous drainage.

Inflation—it refers to the distension with gas especially air.

Inostosis—it refers to the process by which bony tissue is reformed to replace tissue that has been destroyed.

Insidious—unperceived coming on gradually and stealthily.

Intermittent—occurring at intervals with periods of cessation.

Intubation—it refers to the introduction of a tube through the mouth or the nose to allow air, gas or vapor to pass into the lungs.

Iontophoresis—it refers to the therapeutic treatment by electrical introduction of ions into the body tissue.

Ischemia—it refers to the deficiency in the blood supply to a part or an organ which may be due to constriction, contraction or blocking of the arteries.

Isograft—it refers to a graft derived from one member of a pair of monozygotic twins and transplanted to the other.

Jaw winking—it refers to a movement of the lower jaw causing an involuntary movement of the eyelids.

Joint—the place of connection between two bones, allowing of more or less movement on articulation.

Keloid—it refers to a fibrous hyperplastic scar growth on the skin.

Kernicterus—staining of brain tissue caused by accumulation of unconjugated bilirubin in the brain.

Knitting—it refers to the process of repair of a bone fracture.

Lain's disease—burning of the tongue and the soft tissue of the mouth due to electrogalvanism caused by the use of dissimilar metals in dental restoration.

Lancinating—it is the term used to describe shooting, tearing or sharply cutting type of pain.

Leukoplakia—A white patch or plaque that can not be scraped off and can not be characterized clinically or pathologically as any other disease, which is more than 5 mm.

Lesion—a wound or injury or a patch of disease on the skin. A morbid change in tissue function.

Lichen planus—relatively common dermatitis occurs on skin and oral mucous membrane and refers to a lace-like pattern produced by symbiotic algal and fungal colonies on the surface of rocks in nature.

Lipomatosis—it refers to excessive localized accumulation of fats in the tissues.

Ludwig's angina—this condition may be defined as an overwhelming rapidly spreading septic cellulitis involving submandibular, submental and sublingual space bilaterally.

Lymph—it refers to the clear fluid found in the lymphatics vessels.

Lymphadenitis—it refers to the inflammation of the lymph nodes.

Macule—well-circumscribed flat lesion that is noticeable due to the change from the normal skin color to red may be due to inflammation or pigmented due to presence of melanin hemosiderin or other drugs.

Maceration—it refers to the softening of a substance by soaking in a liquid.

Malocclusion—it refers to any deviation from the normal occlusion of the teeth resulting in impaired functions.

Marrow—it refers to the soft tissue canal and interstices of bones.

Marsupialization—it refers to an operation for the evacuation of a cyst and the suturing of its walls to the edges of the wound.

Metastasis—it is defined as spread of tumor by invasion in such a way that discontinuous secondary tumor mass/masses are formed at the site of lodgment.

Metaplasia—it is a reversible change in which one adult cell type is replaced by another adult cell type.

Mesiodens—it refers to supernumerary tooth located at or near the midline in the incisal region of maxilla between the central incisors.

Medicine—it refers to the study and treatment of diseases especially treatment without recourse to surgery or any drug used for the treatment of the disease.

Metabolism—it refers to the physical and chemical changes in the tissue by which a living body is maintained and energy generated.

Mitosis—it is the indirect division of cells, a typical method of cell reproduction.

Mucocele—it is a term used to describe swelling caused by pooling of saliva at the site of injured minor salivary gland.

Muscle—it is a contractile organ by means of which movement is produced in an animal organism.

Muscle spasm—it refers to a sudden involuntary contraction of the muscle or group of muscles attended by pain and interference with function.

Mucus plug—these are incompletely mineralized sialoliths.

Natal teeth—these are teeth which are observed in the oral cavities at birth.

Narcosis—a state of profound unconsciousness or stupor produced by drugs.

Nausea—a feeling of sickness or a tendency to vomit.

Necrosis—it is the sum of the morphologic changes that follow cell death in a living tissue or organs.

Neonatal teeth—these are teeth which erupt during the first 30 days of life.

Neoplasia—it is an abnormal mass of tissue, the growth of which exceeds and is un-coordinate with that of normal tissue and persists in the same excessive manner after cessation of stimuli which evoke the changes.

Neurotropic—attracted to or having an affinity for nervous tissue.

Nevus—it is circumscribed new growth of skin or oral mucosa of congenital origin, presenting as small, elevated, or flat pigmented lesion.

Nodules—this lesion is present deep in the dermis and epidermis and can be moved easily over them.

Nociceptive—relating to any pain producing stimulus, or to pain receptor nerves.

Nosology—it refers to the science of classification of disease.

Numbness—partial or total loss of sensation which may be deliberately induced as in cases of local anesthesia or it may be pathological.

Nutrition—it refers to the process by which food is assimilated.

Ointment—it refers to a fatty semisolid substance used as a base for local medicaments for external application.

Oligodontia—it is agenesis of a few numbers of teeth.

Oncology—it refers to the study of neoplasm.

Operation—anything performed, especially any procedure by a surgeon, either with instruments or by hand.

Oroantral opening—the accidental opening in the floor of maxillary sinus during dental extraction is called as oroantral opening.

Oral submucosa fibrosis—an insidious chronic disease affecting any part of the oral cavity and sometimes the pharynx, preceded by and/or associated with vesicle formation, it is always associated with juxtaepithelial inflammatory reaction followed by fibro-elastic change of the lamina propria, with epithelial atrophy leading to stiffness of the oral mucosa and causing trismus and inability to eat.

Oral medicine—it is that area of dental practice which deals with diagnosis and treatment of oral disease by non-surgical means, which may be localized in the oral cavity or which may be oral manifestation of systemic disease and those phases of dental practice concerned with diagnosis and treatment of medically compromised patients.

Organ—it refers to any separate part of the body having a specific function.

Organism—it refers to any individual plant or animal or an organized body of living cells.

Osteomyelitis—it is an inflammation of bone marrow that produces clinically apparent pus and secondarily affects the calcified component or it is an infection of bone that involves all three components: periosteum, cortex, and marrow or it may be defined as an inflammatory condition of the bone that begins as an infection of the medullary cavity and the Haversian system which extends to involve the periosteum of the affected area.

Papules—these are solid lesions raised above the skin surface that are smaller than 1 cm in diameter.

Pathogenicity—it is the ability of microbial species to produce disease.

Paramolar—it is a supernumerary molar usually small and rudimentary which is situated buccally or lingually to one of the maxillary molars or interproximally between 1st, 2nd and 3rd maxillary molars.

Palpation—it refers to feeling of the affected part by hand.

Pain—it refers to the distressing or unpleasant sensation transmitted by a sensory nerve usually indicative of injury or of disease.

Palliation—it refers to the act of alleviating or affording relief without curing.

Paresthesia—the term refers to perverted sensation like burning, prickling or crawling sensation of the skin.

Parageusia—it refers to an unpleasant taste in the mouth.

Parakeratosis—it refers to any abnormality of the stratum corneum of the epidermis, which may be associated with inflammation of the prickle cell layers causing defective formation of keratin and characterized by the persistence of nuclei.

Paralysis—it is the loss or impairment of muscle function or of sensation due to nerve injury or destruction of neurons.

Pararhizoclasia—it is the inflammatory ulcerative destruction of the deep layers of tissue and the alveolar process about the root of a tooth.

Parenteral—descriptive of methods of drug administration other than by the alimentary canal.

Parodontal—near or next to a tooth sometimes used as synonymous with periodontal.

Parrot tongue—a horny, dry tongue which can not be protruded, seen in typhus and low fever is called as parrot tongue.

Pathogen—any agent that produces or is able to produce disease.

Parulis—it is a mass of granulation tissue which covers the opening of a sinus.

Pathogenesis—the development of disease from its inception to the appearance of characteristic symptoms or lesions.

Pathognomonic—characteristic of one specific disease or pathological condition as distinct from any others.

Pathology—that branch of medicine which is concerned with the structural and functional changes caused by disease.

Pedunculated—in this base of tumor is narrower than the widest part of lesion.

Peridens—supernumerary teeth that are erupted ectopically either buccally or lingually to the normal arch referred to as peridens.

Petechiae—purpuric lesions 1 to 2 mm in diameter.

Permeation—the spreading or extension through tissues or organs, used especially of malignant tumors extending by continuous growth through the lymphatics.

Pericemental abscess—a parodontal abscess not arising from a diseased pulp or an extension of the periodontal pocket.

Percussion—listening to the tapping note with a finger placed on the affected part or in cases of teeth with the help of handle of the probe.

Periodontitis—it is name given to periodontal disease when the superficial inflammation in the gingival tissue extends into the underlying alveolar bone and there is loss of attachment.

Periodontal abscess—an abscess that forms deep in the gums along the tooth root following advanced periodontal disease.

Periodontosis—chronic non-inflammatory destruction of periodontal ligament and the associated alveolar bone.

Phoenix abscess—it is an acute exacerbation of a chronic or suppurative apical periodontitis.

Phlegmon—acute inflammation of the subcutaneous connective tissue.

Pit—it is defined as hollow fovea or indent blind tracts lined with epithelium.

Pilation—it is a hair like fracture found in cranial bones.

Plasmapheresis—it is a method of increasing the number of blood cells in the blood count. From the blood plasma is skimmed out simply on standing and remaining concentrate is reinfused into the patient.

Plaque—it is a solid raised lesion that is over 1cm in diameter.

Pleomorphic—the word pleomorphic means occurring in several distinct shapes.

Pleurodont—having teeth attached to the side of a bony socket or to the side of the jaw.

Plexus—a plexus of nerves or a network of blood or lymphatic vessels.

Pocket—it is an abnormal space developing between the tooth root and the gums.

Poison—any substance that when absorbed into the system of a living body is liable to cause injury and to endanger life.

Poikiloderma—it refers to a combination of atrophy, telangiectasia and pigmentary changes.

Polylophodont—these are teeth with multi-ridged crowns.

Pre-cancerous lesion—morphologically altered tissue in which cancer is more likely to occur than its normal counterpart.

Pre-cancerous condition—it is a generalized state associated with a significantly increased risk of cancer.

Premedication—the administration of drugs or sedatives before treatment, to help in patient management especially with nervous patient.

Prescribe—to write instruction for the preparation, composition and administration of a medicine.

Prevalence—the number of cases of a disease at any given time in any given place.

Priestley's mass—a green or brown stain on the anterior teeth of the young or where reduced enamel epithelium remains over the enamel.

Procheilia—it is the condition in which one lip protrudes forwards of its normal position.

Prognosis—it is the prediction of the course, duration, and termination of the disease and the likelihood of its response to therapy.

Prosthesis—the word prosthesis is used for a manufactured appliance used to take the place of a natural part or to correct a congenital abnormality or it may be defined as an appliance which replaces lost or congenitally missing tissue.

Proteolysis—the process of digestion of proteins and its conversion by enzymes into peptones, proteoses, etc.

Protuberance—the word protuberances refers to a swelling, eminence or knob of the tissue.

Pseudomembrane—it refers to a false membrane, a skin like layer formed by fibrinous exudates containing leukocytes and bacteria.

Pseudoepitheliomatous hyperplasia—in this condition the rete pegs extend far downward, usually accompanied by acanthosis. The cells are normal in size, shape and chromaticity.

Psychosomatic—relating to the mind and the body; particularly relating to the interdependence of mental processes and bodily function.

Pustule—it refers to a raised lesion containing purulent material.

Purpura—it refers to reddish to purple flat lesion caused by blood extravasated from a vessel leaking into subcutaneous tissue.

Pulsation—it is the rhythmic throb or beating as that of the heart.

Pulse—the expansion and contraction of an artery due to increased tension of its walls following contraction of the heart and subsequent relaxation.

Pus—it is a liquid usually yellowish in color formed in certain infection and composed of tissue fluid containing bacteria and leukocytes.

Putrefaction—the decomposition of organic matter through the action of microorganisms, resulting in the production of various solid and liquid compounds and gases giving off a foul odor.

Pyemia—generalized septicemia caused by pyogenic microorganism in the blood stream and marked by the formation of multiple abscesses.

Ranula—the term ranula is used for a mucocele occurring in the floor of mouth in association with ducts of the submandibular or sublingual glands.

Radiology—it is that branch of health sciences dealing with radioactive substances and radiant energy and with the diagnosis and treatment of disease by means of both ionizing (X-rays) and non-ionizing (ultrasound) radiations.

Radiolucent—offering little resistance to X-rays in radiography; almost transparent.

Rash—it refers to a temporary cutaneous eruption.

Recrudescence—the return of symptoms or the recurrence of the disease after a temporary remission.

Recurrence—the return of symptoms or of a disease after a period of remission.

Regurgitation—the return of undigested or partially digested food from the stomach or esophagus to the mouth or of fluid or semifluid to the nose.

Reticular—relating to net or net like structure.

Regeneration—it is replacement of injured tissue by parenchymal cells of the same types.

Retrogenia—it refers to a condition in which chin is set back in relation to the rest of the facial skeleton.

Rh hump—in the deciduous 1st molar crown a characteristic ring like defect may be seen which is called as Rh hump.

Rhinorrhea—it refers to any discharge of fluid from the nose.

Rhizotomy—a surgical division of either a tooth root or a nerve root.

Root dehiscence—it is a pathological condition in which the vestibular surface of the tooth root is exposed to the oral cavity over some or all of the apical two-third of its length.

Rudiment—it refers to an organ or part either imperfectly developed or at an early stage of development.

Rubber jaw—in this condition it is possible to mold the shape of the jaw with the fingers, but teeth will resume its position when the pressure is released.

Satellite abscess—it is a secondary abscess arising from and situated near a primary abscess.

Saburra—it refers to a foul condition of the mouth and teeth or of the stomach due to food debris.

Saucerization—it is the wide and shallow depression occurring about a wound or bone cavity as in osteomyelitis.

Sclerosis—it refers to hardening of vessels or part applied particularly to arteries and to proliferation of connective tissue in the nervous system as a result of degeneration.

Scale—loosened imperfectly cornified parakeratotic superficial layer of skin that is shed as fine, brawny, dirty white, yellowish keratinous dust or large pearly white flakes.

Sequestra—small pieces of necrotic bone which are avascular and which harbor microorganisms are known as sequestra.

Serum—if blood is allowed to clot an amber colored liquid which remains after separation of the clot is known as serum.

Septicemia—the word septicemia implies a overwhelming bacterial proliferation and release of toxins in the blood.

Sessile—it described tumor or growth whose base is widest part of the lesion.

Shock—it is state of inadequate perfusion of all cells and tissues, which at first leads to reversible hypoxic injury, but if sufficiently protracted or grave, to irreversible cell and organ injury and sometimes to the death of the patient.

Sickle cell anemia—in homozygous individuals the whole of HbA (hemoglobin A) is replaced by HbS (hemoglobin S, i.e. an abnormal hemoglobin) and this is known as sickle cell disease.

Sickle cell trait—in heterozygous individuals only 50% of HbA is replaced by HbS and this is known as sickle cell trait.

Sinus—it is a blind tract leading from the surface down to the tissue which is lined by granulation tissue or which may be epithelized.

Sialorrhea (ptyalism)—an increased salivary secretion is termed as sialorrhea or ptyalism.

Sialolithiasis—it is the formation of calcific concretions within parenchyma or ductal system of major or minor salivary glands.

Sinusitis—inflammation of mucosa of paranasal sinuses is referred to as sinusitis. When maxillary sinus is involved it is called as maxillary sinusitis.

Sialoschesis—it is the suppression of the secretion of the salivary glands.

Sign—it defined as any change in the body or its function which is perceptible to a trained observer and may indicate a specific disease.

SLE (systemic lupus erythematosus)—it is characterized by the presence of abnormal serum antibodies and immune complexes.

Slough—it refers to the necrotizing tissue that scales or peels off in ulcerative conditions.

Spongiosis—this term is used to signify intercellular edema of the epithelium, in which intercellular bridges of the stratum spinosum become more prominent.

Stagnation—it refers to the cessation of flow of any circulating fluid in the body.

Sterilization—it is the process of destruction of the microbial life from an article or surface inclusive of bacterial spores.

Sterile abscess—it refers to an abscess containing no microorganisms.

Stenosis—it is the constriction or narrowing of an aperture canal or duct.

Stimulus—it refers to any agent or impulse that excites or promotes a functional reaction.

Stippled—having a mottled or spotted appearance with light and dark patches.

Stomatology—the medical speciality concerned with the mouth and its diseases sometimes used synonymously with dentistry.

Striation—it is a stripe or streak or a series of stripes or streaks.

Stricture—it is an abnormal contraction of any aperture or vessels.

Stridor—it is a harsh whistling sound produced by the respiratory system.

Superficially invasive (micro-invasive) squamous cell carcinoma—a histopathological diagnosis of a routine squamous cell carcinoma, usually well differentiated, which has invaded only slightly into the underlying connective tissues.

Subluxation (hyper-mobility)—it is the unilateral or bilateral positioning of the condyle anterior to the articular eminence, with repositioning to normal to accomplish normal physiologic activity.

Subscription—it is a part of a prescription containing direction for the preparation and compounding of the ingredients of a medicine.

Suzanne's gland—an oral mucous gland found in the alveolo-lingual sulcus near the midline.

Symbiosis—it is the intimate association of two organism of different species.

Symptoms—any indication of the presence or course of a disease either by functional or other changes occurring in the patient.

Syncope—it is a transient loss of consciousness caused by cerebral hypoxia or changes in cerebral blood flow.

Syndesmosis—it is the joining of two bone surfaces by the interposition of connective tissue which forms an interosseous membrane.

Syndrome—a complex of symptoms, occurring together, which characterize one disease or lesion.

Talon's cusp—it projects lingually from the cingulum area of maxillary and mandibular teeth or it is anomalous hyperplasia of cingulum on the lingual of maxillary and mandibular incisor resulting in the formation of a supernumerary cusp.

Taurodontism—body of tooth is enlarge at the expense of root. It is characterized by clinical and anatomical crown of normal shape and size, an elongated body and short root with a longitudinally enlarged pulp chamber.

Tablet—it is a small solid disc containing one dose of a drug.

Tapir mouth—it is a condition characterized by loose thickened lips and caused by atrophy of the orbicularis oris muscle.

Taste—the perception of flavor, a sensation produced by stimulation of the gustatory nerve endings in the tongue with a soluble substance.

Telangiectasia—it is the dilatation of the capillaries and small arteries forming types of angiomas.

Teratoma—it is true neoplasm made up of a number of different types of tissue which are not native to the area in which the tumor occurs.

Thalassemia—it is an inherited impairment of hemoglobin synthesis in which there is partial or complete failure to synthesize a specific type of globin chain.

Therapy—it refers to the treatment of disease.

Thermocautery—it is the use of heat points for cauterization.

Tic—a spasmodic twitching particularly of the facial muscles; a habit spasm.

Tinnitus—the term refers to a ringing noise in the ears.

Toxin—it is a poisonous substance produced by animal or vegetable cells more particularly by bacteria.

Trabeculae—it refers to a septum extending from the outer capsule or envelop into an organ.

Trephine—these are substances prepared by leukocytes from the plasma protein which is necessary for nourishment of tissue cell.

Transplantation—it is the transfer of tissue either from another donor or from one site to another.

Transposition—it is the interchange in position of two adjacent teeth.

Treatment—it is the means used to combat or cure a disease.

Trigonodont—a tooth having three cusps in the form of a triangle.

Trismus—it is the inability to open the mouth because of tonic spasm of the jaw muscles.

Tropics—it is that portion of the surface of the globe where the sun passes directly overhead.

Tumor—it is an autonomous new growth of tissue or it is an abnormal mass of tissue the growth of which exceeds and is uncoordinated with that of normal tissue and persists in the same excessive manner even after the cessation of stimuli which evoked changes.

Tubercle—it is a rounded eminence on bone.

Tumefaction—it is the state of being or becoming swollen.

Twining—it indicates the cleavage of tooth germ into two complete resulting in formation of supernumerary teeth that is mirror image or near image of tooth from which it has developed.

Tyndallization—a method of sterilizing culture media by exposure to steam at 100°C on three successive days for—about 30 minutes each day.

Ulcer—deep crater that extends through the entire thickness of the surface epithelium and involve the underlying connective tissue or defect in epithelium, it is a well circumscribed depressed lesion over which epidermal layer is lost.

Unfavorable fracture—if the associated muscle tends to pull the fragment of the fracture, it is described as unfavorable.

Uvuloptosis—it is a relaxed dropped position of palatine uvula.

Vaccine—it is any material used for preventive inoculation against a specific disease.

Vallate—having a surrounding wall or rim.

Varicosity—it refers to a distended vein, superficial, bluish and painless.

Vasodilator—the term refers to a nerve or some external agent which cause vascular dilatation.

Vesicle—these are elevated blisters containing clear fluid that are under 1 cm in diameter.

Vertigo—it is a sensation of loss of equilibrium in which sufferers feel either that the world is revolving round them or that they are revolving in space.

Verrucous—in this tumor exhibits numerous surface projection.

Virulence—it is a term applied to the properties in a particular strain of microorganism.

Vicarious—relating to a normal process occurring in an abnormal position or under abnormal conditions.

Virus—a complex organic particle of submicroscopic dimensions capable of growth and reproduction only within the cells of the host organism it infects.

Wasting disease of teeth—it is defined as any gradual loss of tooth substance characterized by the formation of smooth polished surfaces without ragged to the possible mechanism of this loss.

Wandering teeth—it is movement of unerupted teeth for no apparent reasons (distal drift).

Wandering abscess—an abscess that tracks through the tissue and finally comes to a point some distance from the original site.

Weal—it is a reddish, raised and circumscribed lesion on the skin, generally caused by a blow or a bite.

Working diagnosis—following reappraisal of diagnostic data at hand including those of follow-up examination which may be seen necessary and which may provide new relevant finding and indicating result from any additional diagnostic procedure.

Wound—it is any injury to the tissues or organs caused by cut, stab or tear, usually going deeper than the outer skin or integument.

Wolf's law—this law states that, the bone structure depends on the strain and stresses to which bone is subjected.

Xerostomia—it is a subjective clinical condition of less than normal amount of saliva.

Xenograft—it is a type of graft derived from a donor of different species.

4

Multiple Choice Questions

1. Hereditary hemorrhagic telangiectasia is seen commonly on:
A. Lips B. Buccal mucosa
C. Tongue D. Palate
2. Raised and pearly white beaded edge is a feature of:
A. Gummatous ulcer B. Rodent ulcer
C. Healing ulcer D. Tuberculous ulcer
3. "Starry sky" appearance is seen in:
A. Pagets disease
B. Cherubism
C. Burkitt's lymphoma
D. Osteomyelitis
4. Delayed type of hypersensitivity reaction to topical antigen is termed as:
A. Stomatitis B. Stomatitis venetana
C. Contact allergy D. Both A and C
5. Kaposi sarcoma is a tumor of:
A. Blood vessels B. Striated muscle
C. Smooth muscle D. None of the above
6. Bednar ulcer is a type of:
A. Traumatic ulcer B. Herpes ulcer
C. Aphthous ulcer D. Allergic ulcer
7. Verocay bodies is associated with:
A. Granular cell myoblastoma
B. Neurilemmoma
C. Neurofibroma
D. Neuralgia
8. Patch test is positive in:
A. Contact allergy
B. Erythema multiforme
C. Aphthous stomatitis
D. Behcet's syndrome
9. All of the above are precancerous condition except:
A. Systemic lupus erythematosus
B. Plummer-Vinson syndrome
C. Lichen planus
D. Syphilis
10. Sutton's disease the ulcer is:
A. Less than 1 cm in diameter and heal without scar
B. Occurs in crops
C. Associated with Beçhet's syndrome
D. More than 1 cm in diameter and heal with scarring
11. Rhabdosarcoma spread by:
A. Local infiltration
B. Lymphatics
C. Blood
D. Direct invasion
12. Bowen disease is:
A. Benign neoplasm of gastrointestinal tract
B. Ulcerative lesion of the skin
C. Vesiculobullous lesion
D. Intra epithelial carcinoma
13. 70-year-old patient comes to the clinic with a tumor of buccal mucosa. The size of the tumor is 3 cm in size with no regional lymph node involvement and no distant metastasis. The TNM stage of the tumor is:
A. T1 N0 M0
B. T2 N0 M0
C. T1 N1 M0
D. T1 N2 M0
14. Radiographic finding of the Pindborg's tumor is:
A. Sunburst appearance
B. Onion peel appearance
C. Driven snow appearance
D. Cherry blossom appearance
15. Ghost cell are seen in:
A. CEOC
B. Ghost teeth
C. AOT
D. OKC
16. The lesion commonly seen in anterior maxilla:
A. Composite compound odontoma
B. Adenomatoid odontogenic cyst
C. Squamous odontogenic tumor
D. All of the above

17. Primary lesion of aphthous stomatitis occurs as:
A. Vesicle B. Bulla
C. Papule D. Blister
18. The cyst with high reoccurrence rate:
A. OKC
B. AOT
C. Globulomaxillary cyst
D. Dentigerous cyst
19. Severe complication of erythema multiforme is called as:
A. Stevens-Johnson syndrome
B. Toxic epidermal necrolysis
C. Both of the above
D. None
20. Ameloblastoma arising from dentigerous cyst is:
A. Mural ameloblastoma
B. Unicystic ameloblastoma
C. Granular ameloblastoma
D. Desmoplastic ameloblastoma
21. Syndrome associated with odontogenic keratocyst is:
A. Gardner's syndrome
B. Gorlin-Goltz syndrome
C. Grinspan syndrome
D. Goldenhar's syndrome
22. Non-inflammatory, non-neoplastic enlargement of the salivary gland is called as:
A. Sialorchitis B. Sialolithiasis
C. Sialadenitis D. Sialosis
23. Odontoma which resembles to the normal teeth is called as:
A. Composite compound odontoma
B. Composite complex odontoma
C. Comminuted odontoma
D. None
24. Multiple odontogenic keratocyst is found in:
A. Marfan's syndrome
B. Ehlers-Danlos syndrome
C. Noonan's syndrome
D. All of the above
25. Focal sclerosing osteomyelitis is associated with:
A. Vital tooth
B. Non-vital tooth
C. Traumatic tooth
D. Cracked tooth
26. Split papule, condyloma latum is seen in:
A. Primary syphilis
B. Secondary syphilis
C. Tertiary syphilis
D. Congenital syphilis
27. Most aggressive type of ameloblastoma, histologically is:
A. Plexiform ameloblastoma
B. Follicular ameloblastoma
C. Basal cell ameloblastoma
D. Granular ameloblastoma
28. All of the following radiographs are taken to diagnose the para nasal sinus pathology *except*:
A. Water's view
B. Caldwell projection
C. Panoramic
D. Submentovertex
29. Leucic glossitis or chronic superficial interstitial glossitis is caused by:
A. Mycobacterium tuberculosis
B. Treponema palladium
C. Actinomycosis
D. Epstein-Barr virus
30. Heerfordt's syndrome is associated with all *except*:
A. Submandibular swelling
B. Uveitis
C. Parotid enlargement
D. Facial palsy
31. Disease causing dermatitis, diarrhea and dementia is:
A. Vit B₁₂ deficiency B. Pellagra
C. Vit B₅ deficiency D. Vit B₆ deficiency
32. A self-healing non-neoplastic inflammatory reaction of salivary gland that mimics a salivary gland neoplasm:
A. Epidermoid carcinoma
B. Pleomorphic adenoma
C. Cystadenoma lymphomatosum
D. Necrotizing sialometaplasia
33. Thrombocytopenic purpura occurs due to:
A. Quantitative defect of platelets
B. Qualitative defects of platelets
C. Both of the above
D. Vit C deficiency
34. Herpes ulcer are seen in:
A. Moveable mucosa
B. Mucosa bound to periosteum
C. Tongue
D. Lips
35. 'Target' or 'bull eye' lesion is typical of:
A. Erythema multiforme
B. Pemphigus
C. Bullous pemphigoid
D. Lichen planus
36. Gingiva is most commonly affected by deficiency of:
A. Vit A B. Vit C
C. Vit D D. Vit B
37. In osteomalacia there is:
A. Defective osteoid + normal mineralization
B. Normal osteoid + defective mineralization
C. Abnormal osteoid + abnormal mineralization
D. Normal osteoid + demineralization
38. The life threatening blood disorder caused due to drugs:
A. Aplastic anemia
B. Pernicious anemia
C. Hemolytic anemia
D. Thrombocytopenia

39. **Hunter's glossitis is seen in:**
 A. Niacin deficiency
 B. Vit V deficiency
 C. Riboflavin deficiency
 D. Vit B₁₂ deficiency
40. **Vesicles or bulla of erythema multiforme, pemphigus vulgaris are:**
 A. Intraepithelial B. Subepithelial
 C. Supraepithelial D. None
41. **Antischkow cells are found in:**
 A. Pemphigus B. Aphthous
 C. Contact allergy D. Stomatitis
42. **Glandular fever is caused due to:**
 A. Varicella-zoster virus
 B. Paramyxovirus
 C. Epstein-Barr virus
 D. Coxsackie A16 virus
43. **Lumbar lordosis is feature of:**
 A. Pellagra B. Osteomalacia
 C. Osteoporosis D. Rickets
44. **Pemphigus is a:**
 A. Inflammatory disease
 B. Autoimmune disease
 C. Skin disease
 D. Noninflammatory disease
45. **The red blood cell in iron deficiency anemia and thalassemia are typically:**
 A. Macrocytic and normochromic
 B. Microcytic and hypochromic
 C. Normocytic and hypochromic
 D. Normocytic and normochromic
46. **Hyperplastic inflammation of the terminal end of the draining sinus is:**
 A. Parulis B. Actinomycosis
 C. Tuberculosis D. Syphilis
47. **Branchial fistula occurs due to fusion of:**
 A. 1st branchial arch with 6th
 B. 1st branchial arch with 2nd
 C. 1st branchial arch with 5th
 D. 2nd branchial arch with 5th
48. **An unusual prominence of premaxilla is seen in:**
 A. Acromegaly B. Paget's disease
 C. Fibrous dysplasia D. Thalassemia
49. **Wash leather slough on the floor of the ulcer is pathognomonic of:**
 A. Tuberculosis ulcer
 B. Aphthous ulcer
 C. Gummatous ulcer
 D. Malignant ulcer
50. **The protein content is less than 4 gm% in:**
 A. Keratocyst
 B. Dentigerous cyst
 C. Radicular cyst
 D. Primordial cyst
51. **Test done for small swelling:**
 A. Patch test
 B. Paget's test
 C. Fluctuation test
 D. Fluid thrill
52. **The most common cause for submaxillary swelling:**
 A. Mumps
 B. Lymphadenopathy
 C. Cellulitis
 D. Periapical abscess
53. **Hemophilia A is due to deficiency of:**
 A. Factor VIII
 B. Factor IX
 C. Factor VII
 D. Platelet deficiency
54. **Swellings that are reducible are:**
 A. Meningocele
 B. Hemangioma
 C. Ranula
 D. Subdermoid cyst
55. **A congenital disease characterized by pancytopenia, bone marrow hypoplasia and congenital anomalies is suggestive of:**
 A. Aplastic anemia
 B. Fanconi anemia
 C. Thrombocytopenia
 D. Pernicious anemia
56. **Pathergy test is diagnostic criteria of:**
 A. Reiters syndrome
 B. Behcet's syndrome
 C. Stevens-Johnson syndrome
 D. Lyell's disease
57. **The temperature that remains above normal throughout the day and fluctuates more than 1 degree in 24 hours is:**
 A. Intermittent fever
 B. Remittent fever
 C. Low grade fever
 D. Continuous fever
58. **Patient suspecting of suffering from infectious mononucleosis which laboratory test should be advised to confirm the diagnosis:**
 A. Dick test
 B. Paul-Bunnell test
 C. Rose Bengal test
 D. Shick test
59. **In sickle cell trait only..... of HbA is replaced by HbS:**
 A. 25%
 B. 50%
 C. 75%
 D. 35%
60. **Gumma is present in:**
 A. Primary syphilis
 B. Secondary syphilis
 C. Tertiary syphilis
 D. Congenital syphilis

61. 'Suttons' disease is other wise commonly called as:
A. Major aphthous stomatitis
B. Minor aphthous stomatitis
C. Recurrent aphthous stomatitis
D. Herpes labialis
62. Ramsay hunt syndrome is caused due to:
A. Coxsackievirus
B. Herpes-zoster virus
C. Herpes simplex
D. Bacterial infection
63. Chvostek's sign is seen in:
A. Hypoparathyroidism
B. Hyperparathyroidism
C. Hypothyroidism
D. Hyperthyroidism
64. Enamel hypoplasia involving incisal edges of the anterior teeth and middle 3rd of deciduous cuspid and 1st molar resulting in a characteristic ring like defect known as 'Rh hump' is characteristic of:
A. Dental fluorosis
B. Erythroblastosis fetalis
C. Turner's syndrome
D. Pernicious anemia
65. Eyes looking toward heaven is a typical appearance seen in:
A. Cherubism
B. Scleroderma
C. SLE
D. Cushing syndrome
66. Mediterranean anemia is commonly known as:
A. Aplastic anemia
B. Thalassemia
C. Pernicious anemia
D. Iron deficiency anemia
67. Bull teeth is seen in:
A. Turner's syndrome
B. Erythema multiforme
C. Taurodontism
D. Dentin dysplasia
68. In geographic tongue there desquamation of:
A. Filiform papillae
B. Fungiform papillae
C. Circumvallate papillae
D. Foliate papillae
69. Kaposi sarcoma is caused by:
A. HSV 2
B. HSV 4
C. HSV 8
D. HTL virus
70. The triad of the mumps consists of epidemic parotitis, orchitis-oophoritis and:
A. Epididymitis
B. Encephalitis
C. Pancreatitis
D. None of the above
71. Cylindroma is:
A. Pindborg's tumor
B. Warthin's tumor
C. Adenoid cystic carcinoma
D. Adenolymphoma
72. 'Lava flowing around the boulders' is seen in:
A. Type 2 dentin dysplasia
B. Type 1 dentin dysplasia
C. Type 3 dentin dysplasia
D. Both A and C
73. Most common leukemia found in children is:
A. ALL
B. CLL
C. AML
D. CML
74. Characteristic appearance of Sjögren's syndrome in sialograms:
A. Cherry blossom
B. Fruitless tree
C. Driven snow
D. Tennis racket
75. Multiple bone involvement, café au lait pigmentation and endocrine disturbance is associated with:
A. Albright's syndrome
B. Jaffe's syndrome
C. Addison's disease
D. Paget's disease
76. Intracellular inclusion bodies detected in herpes simplex infection are called as:
A. Negri bodies
B. Civatte bodies
C. Lipschutz bodies
D. Ruston bodies
77. Leukoplakia type of candidiasis associated with oral epidermoid carcinoma:
A. Acute atrophic
B. Chronic atrophic
C. Chronic hyperplastic
D. Thrush
78. Mikulicz's disease is an:
A. Bacterial infection
B. Viral infection
C. Inflammatory infection
D. Autoimmune disease
79. Cryptococcosis is also known as:
A. Valley disease
B. Torulosis
C. Moniliasis
D. Mucormycosis
80. Id reaction is associated with:
A. Aphthous
B. Herpetic stomatitis
C. Syphilis
D. Candidiasis

81. Christmas disease is due to deficiency of:
 A. Hageman factor
 B. Platelets
 C. Plasma thromboplastin antecedent
 D. Plasma thromplastin component
82. Etiological actor for megaloblastic anemia is:
 A. Vit B₁₂ deficiency
 B. Vit B₆ deficiency
 C. Folate deficiency
 D. Both A and C
83. Candidial infection is seen in all except:
 A. Median rhomboid glossitis
 B. Oral thrush
 C. Geographic tongue
 D. Denture stomatitis
84. Hutchinson's sign is seen in:
 A. Congenital syphilis
 B. Leprosy
 C. Herpes-zoster
 D. Tuberculosis
85. Rigidity of facial muscles producing typical "risus sardonicus" is observed:
 A. Tetanus
 B. MPDS
 C. Bell's palsy
 D. Tetany
86. Most common site for hemangioma tumor is:
 A. Tongue
 B. Lip
 C. Palate
 D. Skin
87. Rubella refers to:
 A. Measles
 B. Smallpox
 C. Chickenpox
 D. German measles
88. Tuberculous infection of submaxillary and cervical lymph node is called as:
 A. Lupus vulgaris
 B. Scrofula
 C. Pott's disease
 D. None of the above
89. Verruciform xanthoma is a variant of:
 A. Histiocytosis-X
 B. Histiocytosis-Y
 C. Eosinophilic granuloma
 D. Achondroplasia
90. Schillings test is done for:
 A. Scarlet fever
 B. Pellagra
 C. Niacin deficiency
 D. B₁₂ deficiency
91. Mumps is caused by:
 A. Paramyxovirus
 B. Pox virus
 C. E-B virus
 D. Orthomyxovirus
92. Cafe' au lait spots are seen in all of the following disease except:
 A. Neurofibromatosis
 B. Fibrous dysplasia
 C. Peutz-Jeghers syndrome
 D. Addison's disease
93. All are the forms of lichen planus except:
 A. Erosive
 B. Atrophic
 C. Hypertrophic
 D. Verrucous
94. Frey's syndrome patient exhibit gustatory sweating and flushing due to damage to:
 A. Chorda tympani nerve
 B. Auriculotemporal nerve
 C. Facial nerve
 D. Facial artery
95. Petrified men is seen in:
 A. MPDS
 B. Myositis ossificans
 C. Trismus
 D. Nasopharyngeal neuralgia
96. Nikolsky's sign is not seen in:
 A. Pemphigus
 B. Hailey-Hailey disease
 C. Epidermolysis bullosa
 D. Lupus erythematosus
97. Hypersensitivity reaction in Steven-Johnson syndrome:
 A. Type 1
 B. Type 2
 C. Type 3
 D. Type 4
98. Koebner phenomenon is seen with:
 A. Erythema multiforme
 B. Discoid lupus erythematosus
 C. Pemphigoid
 D. Lichen planus
99. Gargoyle cells are seen in:
 A. Hurler syndrome
 B. Mucoepidermoid carcinoma
 C. Gingival cyst of adult
 D. Lymphoma
100. Genuiculate neuralgia occurs due to involvement of nerve:
 A. VII
 B. V
 C. X
 D. IX
101. Mumps are characterized as:
 A. Acute non-suppurative sialadenitis
 B. Acute suppurative sialadenitis
 C. Chronic non-suppurative sialadenitis
 D. Chronic suppurative sialadenitis
102. Osteopetrosis shows lab findings:
 A. Increased serum alkaline phosphatase
 B. Decreased serum calcium level
 C. Increased serum acid phosphatase
 D. Decreased serum alkaline phosphatase

- 103. Xerostomia is seen:**
A. Sjögren's syndrome
B. Patient on anticholinergic drugs
C. Patient on radiation therapy
D. All of the above
- 104. Lesion arising due to rupture of salivary duct:**
A. Ranula
B. Mucocele
C. Sialosis
D. Atresia
- 105. Blue sclera, brittle bone opalescent dentin is a sign and symptoms suggestive of:**
A. Paget's disease
B. Osteogenesis imperfecta
C. Marfan's syndrome
D. Osteopetrosis
- 106. Hailey-Hailey disease shows characteristic histological feature:**
A. Basilar hypertrophy
B. No basement membrane
C. Dilapidated brick wall effect
D. Coagulative necrosis
- 107. Pharyngeal pain, dysphagia, sore throat, glossodynia due to elongated styloid process is feature of:**
A. Horner's syndrome
B. Fothergill's disease
C. Frey's syndrome
D. Eagle's syndrome
- 108. Ground glass appearance is a radiographic feature of:**
A. Fibrous dysplasia
B. Hyperparathyroidism
C. Both of the above
D. None of the above
- 109. The gland that can be palpated bimanually is:**
A. Submandibular
B. Sublingual
C. Parotid
D. Sublingual and parotid both
- 110. Serum alkaline phosphatase is increased to 250 Bodansky units in:**
A. Fibrous dysplasia
B. Paget's diseases
C. Osteopetrosis
D. Cherubism
- 111. Triad of Behçet's syndrome consists of all except:**
A. Recurrent oral ulcer
B. Arthritis
C. Recurrent genital ulcers
D. Eye lesions
- 112. Most common neoplasm of major and minor salivary gland:**
A. Warthin's tumor
B. Mucoepidermoid carcinoma
C. Necrotizing metaplasia
D. Cylindroma
- 113. Histologically pemphigus shows:**
A. Acanthosis
B. Acantholysis
C. Keratin pearl
D. Civatte bodies
- 114. Cherubism is a:**
A. Bilateral lesion
B. Unilateral lesion
C. Shows early eruption of permanent teeth
D. Seen at the age of 20-25 years
- 115. Post-cricoid's carcinoma is associated with:**
A. Pernicious anemia
B. Iron deficiency anemia
C. B₁₂ deficiency
D. Aplastic anemia
- 116. von Willebrand's disease shows:**
A. Prolonged bleeding time, normal clotting time
B. Normal bleeding, prolonged clotting time
C. Prolonged bleeding and prolonged clotting time
D. Normal bleeding and normal clotting time
- 117. Biopsy of intact bullae and vesicles done in pemphigus should be of:**
A. Not less than 6 hr
B. More than 12 hr
C. Less than 24 hr
D. More than 48 hr
- 118. Necrotizing sialometaplasia a benign inflammatory lesion occurs most commonly in the sites:**
A. Buccal mucosa
B. Palate
C. Tongue
D. Gingiva
- 119. Patient shows the clinical picture of enlarged both maxilla and mandible with biochemical findings showing increased alkaline phosphatase level and radiographs reveals cotton wool appearance. The most probable diagnosis is:**
A. Monostotic fibrous dysplasia
B. Osteogenic imperfecta
C. Paget's disease
D. Leontiasis ossea
- 120. Localized form of epidermolysis bullosa showing recurrent bullous eruption of hands and feet is seen in:**
A. Herlitz's disease
B. Weber-Cockayne syndrome
C. Epidermolysis bullosa acquista
D. Epidermolysis bullosa latalis
- 121. Nikolsky's phenomenon is a pathognomic sign of:**
A. Pemphigus vulgaris
B. Epidermolysis bullosa
C. Bullosa pemphigus
D. Both A and B
- 122. Epidermolysis bullosa simplex is:**
A. Autoimmune disease
B. Autosomal dominant
C. Autosomal recessive
D. X-linked disease

- 123. Brazilian pemphigus is a mild endemic form of:**
 A. Pemphigus erythematous
 B. Pemphigus vegetans
 C. Pemphigus vulgaris
 D. Pemphigus foliaceus
- 124. Halitosis is caused by all except:**
 A. Periodontal disease
 B. Diabetes
 C. Decayed tooth
 D. Hypertension/Hemangioma
- 125. Irregularly irregular pulse is seen in:**
 A. Atrial fibrillation
 B. Ventricular fibrillation
 C. Ventricular failure
 D. Patent ductus arteriosus
- 126. Clubbing a bullous enlargement of soft part of terminal phalanges of nail positive in:**
 A. Infective endocarditis
 B. Hypothyroidism
 C. Left ventricular failure
 D. None of above
- 127. Life saving drug of choice for the management of Steven-Johnson syndrome:**
 A. Azathioprine
 B. Antiviral drugs
 C. Dapsone
 D. Corticosteroids
- 128. Ramsay hunt syndrome is associated with complication of:**
 A. Shingles
 B. Varicella zoster
 C. Erythema multiforme
 D. Herpangina
- 129. Epidemic disease causing ulcerative lesion restricted to posterior portion of pharynx:**
 A. Herpes simplex
 B. Shingles
 C. Herpangina
 D. Acute lymphonodular pharyngitis
- 130. Intestinal polyp is mainly found in:**
 A. Addison's disease
 B. Peutz-Jeghers syndrome
 C. Hemochromatosis
 D. Neurofibromatosis
- 131. Pathological wearing away of the tooth substance through abnormal mechanical process called as:**
 A. Attrition
 B. Erosion
 C. Abrasion
 D. Abfraction
- 132. Root caries involves commonly:**
 A. Enamel and dentine
 B. Enamel, dentin and pulp
 C. Cementum and dentin
 D. Dentin and pulp
- 133. Stress lesion is another name for:**
 A. Attrition
 B. Abrasion
 C. Erosion
 D. Abfraction
- 134. Bronze diabetes is commonly called as:**
 A. Weber syndrome
 B. Gardener syndrome
 C. Addison's disease
 D. Hemochromatosis
- 135. Pink spot in the roots of the tooth is seen in:**
 A. Internal resorption
 B. Acute pulpitis
 C. Dentinal sclerosis
 D. External resorption
- 136. All of the muscles of the tongue is supplied by hypoglossal except:**
 A. Genioglossus
 B. Hypoglossal
 C. Styloglossus
 D. Palatoglossal
- 137. Deposition of excessive amount of the cementum on the root surface is known as:**
 A. Hypercementosis
 B. Cementicles
 C. Cementum hyperplasia
 D. Both A and C
- 138. Liver cirrhosis, diabetes, cardiac failure and bronze skin are the tetrad mainly seen in:**
 A. Hemochromatosis
 B. Neurofibromatosis
 C. Chronic adrenal insufficiency
 D. Hepatitis
- 139. Amalgam tattoo is an example of:**
 A. Accidental pigmentation
 B. Iatrogenic pigmentation
 C. Endogenous pigmentation
 D. Pigmentation due to drugs
- 140. In order to detect caries radiographically, net mineral loss of the tooth should be:**
 A. Exceed 20 to 30%
 B. Upto 10-20%
 C. More than 40%
 D. 30 to 40%
- 141. Transparent dentin is the term used for:**
 A. Secondary dentin
 B. Dentinal sclerosis
 C. Irregular dentin
 D. Tertiary dentin
- 142. Benign migratory glossitis is called:**
 A. Geographic tongue
 B. Wandering tongue
 C. Glossitis areata exfoliativa
 D. Erythema migrans
 E. All of the above

143. Zone of dentinal sclerosis characterized by deposition of the calcium salt in dentinal tubules in dentin caries is identified as:
A. Zone 1
B. Zone 2
C. Zone 3
D. Zone 4
144. Plumbism is due to:
A. Mercury poisoning
B. Lead poisoning
C. Bismuth poisoning
D. Silver poisoning
145. Dental caries is mainly caused due to:
A. Streptococcus mutans
B. Actinomyces viscosus
C. L. acidophilus
D. Staphylococcus
146. Paper test is done:
A. Allergy
B. Pigmentation
C. Swelling
D. Vit deficiency
147. Cupping in dentin of the tooth is due to:
A. Abrasive lesion
B. Attrition
C. Abfraction
D. Erosive lesion
148. Glossopharyngeal nerve is the nerve for:
A. General sensation for the anterior 2/3rd of the tongue
B. Special sensation for anterior 2/3rd of the tongue
C. General sensation for posterior 1/3rd
D. Special sensation for posterior 1/3rd
149. Cleft palate is associated with all the syndrome except:
A. Pierre-Robin syndrome
B. Down's syndrome
C. Apert's syndrome
D. Ascher's syndrome
150. Which of the radiographs are indicated for the diagnosis of the proximal caries?
A. IOPA
B. Bite wing radiographs
C. Occlusal
D. OPG
151. Bluish green pigmentation on gingiva and teeth called Clapton line is caused due to:
A. Lead
B. Copper
C. Chromium
D. Mercury
152. pH of the dental plaque is critical threshold for demineralization to create dental caries is:
A. pH 5.4
B. pH 5.5
C. pH 5.6
D. pH 5.7
153. Regressive changes that occurs in the teeth that starts from inner surface to outer of the teeth:
A. Abfraction
B. Attrition
C. Erosion
D. Internal resorption
154. Extensibility of the tongue to touch the lip of the nose is termed as:
A. Battle's sign
B. Gorlin's sign
C. Chvostek's sign
D. Bon-bon sign
155. Swift is commonly known as:
A. Silver toxicity
B. Bismuth toxicity
C. Mercurial toxicity
D. Lead toxicity
156. All of the caries are well detected radiographically except:
A. Incipient caries
B. Moderate caries
C. Advanced caries
D. Severe caries
157. Thyroid gland in the embryo form ventral floor of pharynx by means of:
A. Ectodermal evagination of the diverticulum's
B. Endodermal evagination of the diverticulum's
C. Ectodermal invagination of the diverticulum's
D. Endodermal invagination of the diverticulum's
158. Fissured tongue has all other following terms except:
A. Scrotal tongue
B. Lingua villosa
C. Plicated tongue
D. Lingua dissecta
159. Pierre-Robin syndrome is the combination of the disease except:
A. Micrognathia
B. Cleft palate
C. Glossoptosis
D. Macroglossia
160. Systemic disease not causing gingival enlargement:
A. Leukemia
B. Crohn's disease
C. Orofacial angiomatosis
D. Diabetes
161. Greenstick fracture are seen in:
A. Newborn
B. Children
C. Youngsters
D. Adult
162. Sharpened pencil appearance of the condyle is seen in:
A. Osteoarthritis
B. Rheumatic fever
C. Rheumatoid arthritis
D. Osteoporosis

163. Maxillary sinus is best visualized by:
 A. Periapical radiographs
 B. Waters view
 C. Submentovertex
 D. Lateral skull view
164. Physical medication used in management of MPDS are all *except*:
 A. Oral myofunctional therapy
 B. Diathermy and ultrasound
 C. Electrical stimulation
 D. Voluntary resistance
165. Stiff joint is term used for:
 A. Internal derangement
 B. Ankylosing spondylitis
 C. Ankylosis
 D. Infective arthritis
166. In edentulous patient bilateral condylar fracture is treated by:
 A. Using gunning splint
 B. Arch bar
 C. Arch wire
 D. Only immobilization in normal position
167. Subcutaneous nodules mainly seen in:
 A. Psoriatic arthritis
 B. Rheumatic arthritis
 C. Osteoarthritis
 D. Klinefelter syndrome
168. Minute areas of degeneration filled with fibrous tissues just below the bony surface of the condyle in osteoarthritis called as:
 A. Lipping of the bone
 B. Ely's cyst
 C. Osteophyte
 D. Eburnation
169. Good transillumination of sinus indicates:
 A. Presence of pus in sinus
 B. Presence of solid lesion in sinus
 C. Mucosal thickening of the sinus
 D. Presence of air filled sinus
170. Malignant tumor of the cartilage:
 A. Osteochondroma
 B. Chondrosarcoma
 C. Synovial chondromatosis
 D. Osteoma
171. Laskin's cardinal sign involves the following features except:
 A. Unilateral pain in preauricular area
 B. Muscle tenderness
 C. Masticatory muscle spasm
 D. Limited jaw movement
172. Irregular shaped reddish area of depapillation and thinning of the dorsal tongue epithelium surrounded by the zone of regenerative papillae is shown in:
 A. Fissured tongue
 B. Median rhomboid glossitis
 C. Benign migratory glossitis
 D. Hairy tongue
173. Color of the pigmentation caused due to hemosiderin is:
 A. Blue
 B. Brown
 C. Grey
 D. Black
174. Dental caries that is difficult to differentiate radiographically:
 A. Occlusal caries
 B. Buccal caries
 C. Mesial caries
 D. Distal caries
175. Failure to unite palatal process due to disturbances in normal fusion of the palatal shelves causes:
 A. Only cleft of hard palate
 B. Both cleft of hard and soft palate
 C. Cleft of lip, hard and soft palate
 D. Cleft of hard, soft palate and uvula
176. Phlebectasia is the term used for:
 A. Lingual varices
 B. Varicosity
 C. Both A and B
 D. None of the above
177. Fibrotic gingival enlargement is seen in all except:
 A. Due to drug cyclosporine
 B. Due to nifedipine
 C. Granuloma pyogenicum
 D. Idiopathic
178. Parade ground fracture involves:
 A. Unilateral condylar fracture
 B. Bilateral condylar fracture
 C. Mandibular body fracture
 D. Ramus and angle of mandibular fracture
179. Corticosteroids are contraindicated in:
 A. Herpes simplex
 B. Mandibular osteoarthritis
 C. Submucous fibrosis
 D. Lichen planus
180. Antrum of highmore is the term used for:
 A. Sphenoid sinus
 B. Cavernous sinus
 C. Maxillary sinus
 D. Ethmoidal sinus
181. Gout is characterized with following except:
 A. Joint pain
 B. Swelling over the joint
 C. Elevated blood uric acid
 D. Multiple bone fracture
182. Beauty vitamin is another name for:
 A. Vit B₁
 B. Vit B₆
 C. Vit B₁₂
 D. Vit B₂
183. Macrognathia seen in:
 A. Robin's syndrome
 B. Turner's syndrome
 C. Leontiasis ossea
 D. Marfan's syndrome

184. Electrical burns causes tissues necrosis is characterized by:
A. Coagulation of the protein
B. Destruction of the neural tissues
C. Liquefaction of the fat
D. Vaporization of the tissues fluids
185. Dislocation and disarticulation of the teeth which is abnormally mobile and are displaced called as:
A. Concussion
B. Avulsion
C. Luxation
D. Contusion
186. Dry socket is referred to:
A. Postoperative osteitis
B. Alveolalgia
C. Focal osteomyelitis
D. All of the above
187. Facial hemiatrophy is known as:
A. Marfan's syndrome
B. Parry-Romberg syndrome
C. Hallerman-Steriff syndrome
D. Crouzon's syndrome
188. Gummy smile seen mostly in case of:
A. Friedreich disease
B. Macrognathia
C. Micrognathia
D. Mandibulofacial dystosis
189. Diabetes is associated with:
A. Hand-Schuller-Christian
B. Lichen planus
C. None of the above
D. Both A and B
190. Blow out fracture is due:
A. Calcification in the sinus
B. Increased in intraorbital pressure
C. Trauma to the chin
D. All of the above
191. Taurodontism is associated with:
A. Crouzon;s syndrome
B. Klinefelter's syndrome
C. Binder syndrome
D. Treacher-Collins syndrome
192. Inflammation of the mucosa of paranasal sinus is referred as:
A. Pansinusitis
B. Sinusitis
C. Antroliths
D. Maxillary sinusitis
193. Internal derangement occurs due to trauma from malocclusion results in the spasm of:
A. Temporalis
B. Lateral pterygoid
C. Medial pterygoid
D. Masseter
194. Macroglossia can be result of:
A. Lymphangioma
B. Neurofibroma
C. Lipoid proteinosis
D. All of the above
195. In unilateral ankylosis patient's face is deviated towards:
A. Affected side
B. Unaffected side
C. Undeviated
D. None of the above
196. Erosion of the bone found in which stage of the rheumatoid arthritis:
A. Stage I
B. Stage II
C. Stage III
D. Stage IV
197. The gingival fibers that helps to maintain the position of free gingiva:
A. Dento-gingival
B. Alveola gingival
C. Circular fibers
D. Transseptal fibers
198. ANUG is caused by:
A. Only spirochetes
B. Both fusiform bacilli and spirochetes
C. Fusiform bacilli, spirochetes and bacteroid intermedius
D. Spirochetes and bacteriod intermedius
199. Carcinoma of the tip of the tongue is drained in:
A. Submental lymph nodes
B. Jugulo-omohyoid node
C. Submandibular node
D. Both A and B
200. Volkmann's chelitis is most severe suppurative form of:
A. Granulomatous chelitis
B. Angular chelitis
C. Actinic chelitis
D. Glandular chelitis
201. 'Rule of ten' is used for the management of cleft lip involves the following criteria *except*:
A. Age
B. Body weight
C. Body fluid
D. Hb%
202. Miescher's syndrome is also known as:
A. Glandular chelitis
B. Granulomatous chelitis
C. Plasma cell chelitis
D. Exfoliative chelitis
203. A patient with Bell's palsy has.....palsy:
A. Upper motor neuron nerve
B. Upper motor neuron VII nerve
C. Lower motor neuron V nerve
D. Lower motor neuron VII nerve

204. Characteristic 'tram line' calcifications in the skull radiographs is observed in:
 A. Cleidocranial dysostosis
 B. Sturge-Weber syndrome
 C. Paget's disease
 D. Maccune-Albright syndrome
205. Which of the following appears to arise from the sunlight?:
 A. Basal cell carcinoma B. Ewing's sarcoma
 C. Kaposi sarcoma D. Verruca vulgaris
206. A hamartoma is:
 A. Any collection of the blood clot
 B. A hemorrhagic cyst of the thigh
 C. Developmental malformation
 D. A tumor of soft tissues
207. 'Rubber man' appearance seen in:
 A. Marfan's syndrome
 B. Gorham syndrome
 C. Ehlers-Danlos syndrome
 D. First arch syndrome
208. Human immunodeficiency virus binds to macrophages having:
 A. CD4 molecule
 B. CD4 and CXCR molecule
 C. CD4 and CCR5
 D. CXCR and CCR5 molecule
209. Which of the following conditions is the most common complication of radioiodine treatment of Grave's disease?:
 A. Thyroid storm B. Subacute thyroiditis
 C. Thyroid cancer D. Hypothyroidism
210. Microscopic appearance of the geographic tongue resemble:
 A. Lichen planus
 B. Lupus erythematosus
 C. Erythema multiforme
 D. Psoriasis
211. Which of the following test is most specific test to diagnose syphilis?
 A. VDRL test B. Wassermann test
 C. RPR test D. FTA-ABS
212. Hemochromatosis presents with all except:
 A. Micronodular cirrhosis
 B. Diabetes mellitus
 C. Skin pigmentation
 D. Hepatitis
213. Strawberry tongue is seen in:
 A. Scarlet fever
 B. Wegner's granulomatosis
 C. Midline lethal granuloma
 D. Sarcoidosis
214. Among the following pain theories, the most accepted theory is:
 A. Specificity theory
 B. Pattern theory
 C. Gate control theory
 D. Calcium displacement theory
215. Definite loss of lamina dura is seen in:
 A. Secondary hypercemia
 B. Hyperparathyroidism
 C. Primary hyperthyroidism
 D. Secondary hypothyroidism
216. Treatment of choice for mixed parotid tumor is:
 A. Enucleation
 B. Superficial parotidectomy
 C. Radical parotidectomy
 D. Radiation
217. Which one given below is a DNA virus:
 A. Poliovirus B. Adenovirus
 C. Astor viridae D. Hepatitis-A virus
218. The Philadelphia chromosome is a feature of:
 A. Luekemoid reaction
 B. Chronic myeloid leukemia
 C. Acute lymphoblastic leukemia
 D. Chronic lymphocytic leukemia
219. Ranula is a:
 A. Retention cyst B. Implantation
 C. Begin tumor D. Malignant tumor
220. Failure of descent of thyroid analouge can be seen in the tongue:
 A. In anterior 2/3rd of dorsal aspect
 B. In posterior 1/3rd of dorsal aspect
 C. Near the base of the tongue close to foramen caecum
 D. In anterior 2/3rd of inferior surface
221. Most common salivary gland tumor arising in the jaws:
 A. Pleomorphic adenoma
 B. Adenoid cystic carcinoma
 C. Mucoepidermoid carcinoma
 D. Acinic cell carcinoma
222. Kussmaul's respiration occurs in response to:
 A. Decrease in pH of the blood
 B. Increase in pH of the blood
 C. Obstructive pulmonary disease
 D. Carbon monoxide poisoning
223. Enamel hypoplasia occurring from the local infection is called:
 A. Amelogenesis dysplasia
 B. Plumbism
 C. Dental fluorosis
 D. Turner's teeth
224. The absence of clavicle are seen in:
 A. Cleidocranial dysplasia
 B. Osteogenesis imperfecta
 C. Sturge-Weber anomaly
 D. Idiopathic bone cavities
225. The stabbing nature of the pain in trigeminal neuralgia mimic the pain caused by:
 A. A cracked tooth
 B. Acute reversible pulpitis
 C. Acute irreversible pulpitis
 D. Acute apical periodontitis

226. Sialoliths are the stones found in salivary duct and duct is mainly composed of:
A. Hydroxyapatite
B. Potassium chloride
C. Unknown compounds of phosphate
D. Calcium chloride
227. 'Denture sore mouth' are also known as:
A. Acute pseudomembranous candidiasis
B. Acute atrophic candidiasis
C. Chronic atrophic candidiasis
D. Chronic hyperplastic candidiasis
228. The virus responsible to cause Hodgkin's lymphoma:
A. Herpes virus
B. E-B virus
C. Coxsackievirus
D. HHV6 virus
229. Horton's syndrome is:
A. Trigeminal neuralgia
B. Sphenopalatine neuralgia
C. Parkinsonism
D. None
230. Shingles of the geniculate ganglion with ear eruption and facial paralysis is termed as:
A. Peutz-Jeghers syndrome
B. Melkersson-Rosenthal syndrome
C. Ramsay Hunt syndrome
D. Gardner's syndrome
231. Hot potato voice is characteristically seen in:
A. Pterygomandibular space infection
B. Retropharyngeal space infection
C. Pretracheal space infection
D. Lateral pharyngeal space infection
232. Acute pyogenic infection produces:
A. Leukopenia
B. Leukocytosis
C. Neutropenia
D. Lymphopenia
233. Hanging drop sign is best seen in:
A. Water's projection
B. Orthopantomogram
C. Submentovertex
D. Occlusal view
234. A fractured mandibular condyle is displaced by the action of:
A. Temporalis
B. Masseter
C. Lateral pterygoid
D. Medial pterygoid
235. In hemolytic anemia the urobilinogen level in the blood would:
A. Increase
B. Absent
C. Decrease
D. Mildly decrease
236. Epstein-Barr virus has been implicated in the etiology of:
A. Oral Lichen planus
B. Oral hairy leukoplakia
C. Oral hairy black tongue
D. Oral melanoplakia
237. A 20 years female came to the clinic with severe pain and redness over the dorsum of the foot. Past history of severe abdominal pain episodes were present. Peripheral smear showed anemia with presence of poikilocytes your likely diagnosis is:
A. Hemoglobin C disease
B. Thalassemia
C. Sickle cell anemia
D. G6PD deficiency
238. In young permanent teeth the best method of sensitivity testing for the traumatized teeth is carbon dioxide snow which can go up to temperature of:
A. -20°C
B. -30°C
C. -40°C
D. -78°C
239. Most common dislocation of the TMJ is:
A. Anterior
B. Lateral
C. Posterior
D. Medial
240. Uniform widening of the PDL at the expense of the adjacent bone occurs in:
A. Langerhan's cell histiocytosis
B. von Recklinghausen's disease
C. Scleroderma
D. Osteoporosis
241. The nerves commonly affected in shingles are C3, T5, L1, L2 and:
A. 1st division of trigeminal nerve
B. 2nd division of trigeminal nerve
C. 3rd division of trigeminal nerve
D. Facial nerve
242. Fibromatosis gingiva is inherited as:
A. Autosomal dominant
B. Autosomal recessive
C. Both A and B
D. None
243. Synonyms used for necrotizing ulcerative gingivitis is all except:
A. Vincent infection
B. Trench mouth
C. Acute ulceromembranous gingivitis
D. Desquamative gingivitis
244. Most common sign of trauma from occlusion:
A. Occlusal facet
B. Drifting of the teeth
C. Increased mobility of the teeth
D. Gingival bleeding

- 245. Rose waaler test is used to diagnose:**
 A. Rheumatic fever
 B. Osteoarthritis
 C. Rheumatoid arthritis
 D. Osteopetrosis
- 246. Reimplantation of tooth is treatment can be done in case of:**
 A. Concussion
 B. Avulsion
 C. Luxation
 D. Extrusion
- 247. Concrecence occurs:**
 A. After root formation
 B. After crown formation
 C. During crown formation
 D. After complete eruption of tooth
- 248. Talon's cusp arises from:**
 A. Palatal surface
 B. Occlusal surface
 C. Labial surface
 D. Cervical region
- 249. Emphysema is due to:**
 A. Presence of gas
 B. Presence of pus
 C. Presence of blood
 D. Presence of watery fluid
- 250. In Ellis and Davey's classification, displacement of tooth without crown or root fracture includes in:**
 A. Class V
 B. Class VI
 C. Class VII
 D. Class VIII
- 251. Fusion involves union of:**
 A. Only crown of teeth
 B. Only roots of tooth
 C. Both crown and roots of teeth
 D. None of the above
- 252. The most common facial fracture is:**
 A. Maxillary fracture
 B. Mandibular fracture
 C. Zygomatic fracture
 D. Nasal bone fracture
- 253. 'Hanging drop appearance' into antrum seen in:**
 A. Zygomatic complex fracture
 B. Blow-out fracture
 C. Sinus contusion
 D. Squamous cell carcinoma of the antrum
- 254. Exuberent development of fourth lobe gives rise to:**
 A. Supernumerary tooth
 B. Enamel pearl
 C. Bolk cusp
 D. Talon's cusp
- 255. Dens in dent' is due to:**
 A. Focal growth retardation
 B. Active proliferation of enamel organ
 C. Increased intravascular pressure
 D. All the above
- 256. Dilaceration seen mostly in:**
 A. Maxillary 1st molar
 B. Mandibular 1st molar
 C. Maxillary incisor
 D. Mandibular incisors
- 257. In unilateral ankylosis:**
 A. Chin retracted towards affected side
 B. Chin retracted towards unaffected side
 C. Chin is in midline position
 D. None of above
- 258. Dens Evaginatus is the term referred to:**
 A. Gestant odontome
 B. Bolk cusp
 C. Leong's premolar
 D. Carabelli's cusp
- 259. Following is the self-reducing incomplete dislocation:**
 A. Anterior dislocation
 B. Subluxation
 C. Central dislocation
 D. Posterior dislocation
- 260. Internal derangement occurs due to:**
 A. Articular surface remodeling
 B. Disc deformation
 C. Displacement of condyle from its position
 D. All of the above
- 261. Enamel pearl occurs most commonly in:**
 A. Trifurcation of maxillary molars
 B. Bifurcation of maxillary premolars
 C. Bifurcation of mandibular molars
 D. Cervical region of incisors
- 262. Dens in dent is also called as:**
 A. Dilated composite odontome
 B. Gestant odontome
 C. Dens invaginatus
 D. All of the above
- 263. Natal teeth may be associated with:**
 A. Paget's disease
 B. Ellis-van Creveld syndrome
 C. Gardener's syndrome
 D. Cleidocranial dysplasia
- 264. Carabelli's cusp is located:**
 A. Mesiopalatally
 B. Distopalatally
 C. Mesiobuccally
 D. Distobuccally
- 265. Which is the most specific test used to eliminate false positive result is:**
 A. ELISA test
 B. Western blot test
 C. Indirect immunofluorescence test
 D. Polymerase chain reaction
- 266. Actinic keratosis commonly seen in:**
 A. Females and black
 B. Males and black
 C. Females and whites
 D. Males and whites

- 267. Rhagades found in:**
A. Angular cheilitis
B. Eczematous cheilitis
C. Contact cheilitis
D. Exfoliative cheilitis
- 268. Punched out lesions occurs in ANUG in:**
A. Stage 2
B. Stage 3
C. Stage 4
D. Stage 1
- 269. In AIDS patient following findings are present except:**
A. Increased number of helper T cells
B. Decreased serum globulin level
C. Decreased ratio of T helper cells to T suppressor cells
D. Increased level of circulating immune complex
- 270. Goldenhar syndrome consists of:**
A. Agenesis of condyle
B. Hyperplasia of condyle
C. Double condyle
D. Condylar hypoplasia
- 271. Osteoarthritis is called as:**
A. Rheumatoid arthritis
B. Degenerative arthritis
C. Psoriatic arthritis
D. All of the above
- 272. Actinic elastosis is:**
A. Solar elastosis
B. Solar cheilosis
C. Actinic keratosis
D. All of the above
- 273. Most common neoplasm associated with AIDS is:**
A. Non-Hodgkin's lymphoma
B. Kaposi sarcoma
C. Hodgkin's lymphoma
D. Squamous cell carcinoma
- 274. Hypoplasia of condyle is not seen in:**
A. Larsen's syndrome
B. Mobius syndrome
C. Oral facial digital syndrome
D. Pierre Robin syndrome
- 275. Blue domed cyst consists of cavity filled with:**
A. Air
B. Blood
C. Watery fluid
D. Pus
- 276. Hemisepta is the bony defect more common on:**
A. Distally
B. Mesially
C. Buccally
D. Lingually
- 277. Most striking feature of juvenile periodontitis is:**
A. Affects incisors and 1st molar
B. Lack of clinical information despite presence of deep pocket
C. Distolabial migration of maxillary incisors
D. All of the above
- 278. Fat soluble vitamin is stored in:**
A. Liver
B. Kidney
C. Colon
D. Gut
- 279. Outward deviation of one of the eye:**
A. Proptosis
B. Strabismus
C. Diplopia
D. None
- 280. Cleidocranial dysplasia is also known as:**
A. Crouzon's disease
B. Friedreich disease
C. Pagets disease
D. Marie and Sainton disease
- 281. Periodontosis is:**
A. Slowly progressive periodontitis
B. Refractory periodontitis
C. Necrotizing ulcerative periodontitis
D. Juvenile periodontitis
- 282. The most common type of odontogenic cyst is:**
A. OKC
B. Dentigerous cyst
C. Primordial cyst
D. CEOC
- 283. Displaced condylar fracture is well demonstrated on:**
A. AP view
B. Lateral projection
C. Both A and B
D. Orthogram
- 284. Gorlin cyst is:**
A. OKC
B. CEOC
C. Radicular cyst
D. Dentigerous cyst
- 285. Fredrich's disease is called as:**
A. Facial hemiatrophy
B. Facial hemihypertrophy
C. Cleidocranial dysplasia
D. Mandibulofacial dysostosis
- 286. Vestigial cyst is:**
A. Median palatal cyst
B. Nasopalatine cyst
C. Nasoalveolar cyst
D. Globulomaxillary cyst
- 287. Down syndrome consists of:**
A. Microdontia
B. Supernumerary teeth
C. Fissured tongue
D. All of the above
- 288. Single tooth germ invaginates resulting in incomplete formation of two teeth having a bifid crown with single root called as:**
A. Tuning
B. Dens invaginatus
C. Concrescence
D. Gemination

289. Delayed eruption of the teeth occurs in all the disease *except*:
- A. Cleidocranial dysplasia
 - B. Rickets
 - C. Cretinism
 - D. Chondroectodermal dysplasia
290. Simple bone cyst is the term used for:
- A. Traumatic bone cyst
 - B. Aneurysmal bone cyst
 - C. Epidermoid cyst
 - D. Globulomaxillary cyst
291. AIDS occurs mostly in:
- A. Homosexual
 - B. Heterosexual
 - C. Intravenous drugs users
 - D. Newborn infants by infected mother
292. Shell teeth appearance is seen radiographically in:
- A. Dentin dysplasia
 - B. Dentinogenesis imperfecta
 - C. Regional odontodysplasia
 - D. Amelogenesis imperfecta
293. Thin white opaque line running across the tooth surface due to fluorosis under:
- A. Score 0
 - B. Score 1
 - C. Score 3
 - D. Score 2
294. Riga fede is a complication of:
- A. Natal teeth
 - B. Distomolar teeth
 - C. Talon cusp
 - D. Peg shaped lateral
295. Snow capped teeth seen in amelogenesis imperfecta due to:
- A. Hypoplasia of enamel
 - B. Hypocalcification of enamel
 - C. Hypomaturation of enamel
 - D. Trauma to tooth
296. Papillion-Lefevre syndrome is inherited as:
- A. Autosomal recessive
 - B. Autosomal dominant
 - C. Autosomal recessive and X-linked
 - D. Autosomal dominant and X-linked
297. Supernumary tooth that erupts ectopically in normal arch is referred to:
- A. Mesiodens
 - B. Para dens
 - C. Peridens
 - D. Distomolar
298. Teeth erupt prematurely during birth is:
- A. Natal teeth
 - B. Neonatal teeth
 - C. Congenital
 - D. All of the above
299. Bird/fish facial appearance occurs commonly in:
- A. Cleidocranial dysplasia
 - B. Paget's disease
 - C. Mandibulofacial dysostosis
 - D. Crouzon's disease
300. The 'prow of boat' appearance occurs in:
- A. Ellis-van Creveld disease
 - B. Polydactylism
 - C. Arhinencephaly
 - D. All of the above

ANSWERS

1 A	2 B	3 C	4 D	5 A	6 A	7 B	8 A	9 A	10 D	11 B
12 D	13 B	14 C	15 A	16 D	17 C	18 A	19 C	20 A	21 B	22 D
23 A	24 D	25 B	26 B	27 D	28 D	29 B	30 A	31 B	32 D	33 B
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